CONGENITAL MALFORMATIONS OF THE HEART

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CONTENTS

VOLUME I GENERAL CONSIDERATIONS

VOLUME II SPECIFIC MALFORMATIONS

Part One
Malformations with a Venous Arterial Shunt

PART TWO

MALFORMATIONS OF THE RIGHT SIDE OF THE HEART (WITH OR WITHOUT A VENOUS ARTERIAL SHUNT)

PART THREE

MALFORMATIONS IN WHICH THE PRIMARY SHUNT IS FROM LEFT TO RIGHT

Part Four

Malformations of the Left Side of the Heart

CONGENITAL MALFORMATIONS OF THE HEART

Volume II



CHAPTER VI

THE TETRALOGY OF FALLOT

PULMONARY STENOSIS OR ATRESIA COMBINED WITH DEXTROPOSITION OF THE AORTA

IN 1671 a case of a tetralogy of Fallot was reported by Nils Stensen[1] This is now considered to be the first report of the malformation Over a hundred years later, in 1777, Sandifort wrote a detailed account of a boy who suffered from a similar condition This report is remarkable for its description of the clinical manifestations and also because the parents requested an autopsy In this instance the boy had been perfectly normal at birth and therefore the physicians attributed the unusually livid color which had been present since one year of age to an acquired disease Sandifort went on to say, So imagine our surprise when we found the heart to be grossly malformed and that the aorta arose in part from the right ventricle a condition which must have been present from birth [3] According to Peacock,[4] in 1783 Hunter reported a case of a child who was always dark-colored and had presented unusual symptoms of malformation of the heart since shortly after birth, and was remarkably thin He was liable to paroxysms of difficulty in breathing but could arrest them by lying on the carpet when they were coming on This, to the author s knowledge, is the first account of an attack of paroxysmal dyspnea, which is so common in patients with a tetralogy of Fallot The first case which came under Peacock s[4] observation was that of a boy who was born healthy and continued to thrive until he was vaccinated at the age of three months By 1866, when Peacock published the second edition of his book on congenital heart disease, he was evidently familiar with this malformation, as the first nine of the eighteen cases of malformation of the heart were cases of pulmonary stenosis or pulmonary atresia combined with dextroposition of the aorta This is clearly shown by his beautiful illustrations

In 1888 Etienne Fallot made a detailed report of a series of cases with pulmonary stenosis or atresia and clarified the syndrome which now bears his name The condition, as originally described by Fallot, consists of four features *pulmonary stenosis or atresia* combined with *dextroposition of the aorta a ventricu*

right ventricle Embryologically the pulmonary artery and the outflow tract of the right ventricle are both derived from the bulbus cordis [8] For this reason the underdevelopment of the pulmonary artery is frequently associated with narrowing, or stenosis, of the outflow tract of the right ventricle

The usual type of infundibular stenosis is illustrated in Figure VI–1 Drawing A shows the exterior surface of the right ventricle for orientation, drawing B shows an infundibular stenosis and a small infundibular chamber, and drawing C shows the manner in which the aorta over rides the right ventricle In this specimen, as is usual, the aorta is considerably larger than the pulmonary artery and the pulmonary valve, though small, is of normal structure Occasionally the stenosis is of the valvular type but even in such instances there is usually some degree of infundibular stenosis In rare instances the stenosis may be entirely infundibular and the pulmonary valve of normal size and structure, as illustrated in Figure VI–2 Occasionally the pulmonary stenosis is purely valvular In every instance, regardless of the location or the severity of the pulmonary stenosis, the pulmonary artery arises from the right ventricle

The structure of the heart with a diffuse narrowing of the outflow tract of the right ventricle is illustrated in Figure VI–3 Although the pulmonary stenosis was extreme, this child lived for three years and died of a subacute bacterial endocarditis Figure VI–4 illustrates a tetralogy of Fallot with pulmonary atresia A comparison of Figures VI–3 and VI–4 shows how slight may be the difference between a tetralogy of Fallot with pulmonary stenosis and one with pulmonary atresia Nevertheless, in the former case the ductus arteriosus was obliterated and the blood from the right ventricle was pumped through the stenosed pulmonary artery to the lungs, whereas in the latter the circulation to the lungs was established by way of the ductus arteriosus It is important to emphasize that in a tetralogy of Fallot with pulmonary atresia the ductus arteriosus, even though it is essential for life, only rarely remains patent

The pulmonary stenosis, either infundibular or valvular, may be so severe that there is but a pinpoint opening Furthermore, in some instances a deposit of calcareous material is found at autopsy, either on the stenotic valve or along the distal margin of the infundibular obstruction Such concretions, which are almost certainly deposited after birth, cause further narrowing of the pulmonary orifice In some instances it seems as if the abnormal area grows less rapidly than the normal part and thus the pulmonary stenosis becomes proportionally greater In yet other instances the musculature of the infundibular region becomes progressively more hypertrophied, which in turn increases the obstruction Conse

lar septal defect, and *hypertrophy of the right ventricle* hence the name "tetralogy"

Pulmonary stenosis or pulmonary atresia, when combined with dextroposition of the aorta, was classified by Fallot as the same anomaly Although anatomically the two conditions are distinct, it is reasonable to classify them together because there are all grades of pulmonary stenosis As the pulmonary stenosis increases in severity, the obstruction becomes so great that there is a functional pulmonary atresia Nevertheless, the two conditions differ from each other in one important respect, namely the pathway by which the blood reaches the lungs In patients with pulmonary stenosis the circulation to the lungs, although reduced, follows its normal course it passes from the right ventricle through the stenosed pulmonary orifice to the pulmonary artery and thence to the lungs Consequently a tetralogy of Fallot with pulmonary stenosis, especially if the stenosis is not extremely severe, may be compatible with life for many years Indeed, it was long considered the commonest malformation of the cyanotic type compatible with relative longevity In contrast to this, when there is pulmonary atresia or an extremely severe degree of pulmonary stenosis, the circulation to the lungs is by way of the ductus arteriosus Consequently the infant can survive the closure of the ductus arteriosus only if the collateral circulation develops with great rapidity This rarely occurs For this reason most infants with a tetralogy of Fallot and pulmonary atresia die at an early age

Spitzer[6] has classified the tetralogy of Fallot as a Type I transposition, because of the partial rotation of the great vessels The rotation, however, concerns only the aorta, the aorta over rides the ventricular septum and arises in part from the right ventricle In the classic case of a tetralogy of Fallot, the aorta only slightly over rides the ventricular septum In other instances the aorta arises from the right ventricle to such an extent that it is virtually transposed The pulmonary artery, however, occupies its normal position, therefore, although the aorta is rotated on its axis, the great vessels are not transposed

NATURE OF THE MALFORMATION

As previously mentioned, the four features which constitute the tetralogy of Fallot are pulmonary stenosis, dextroposition of the aorta, a ventricular septal defect, and right ventricular hypertrophy The ductus arteriosus undergoes normal obliteration, the foramen ovale is closed

The pulmonary stenosis is usually of the infundibular type, generally the abnormality involves both the pulmonary artery and the outflow tract of the

right ventricle Embryologically the pulmonary artery and the outflow tract of the right ventricle are both derived from the bulbus cordis [8] For this reason the underdevelopment of the pulmonary artery is frequently associated with narrowing or stenosis, of the outflow tract of the right ventricle

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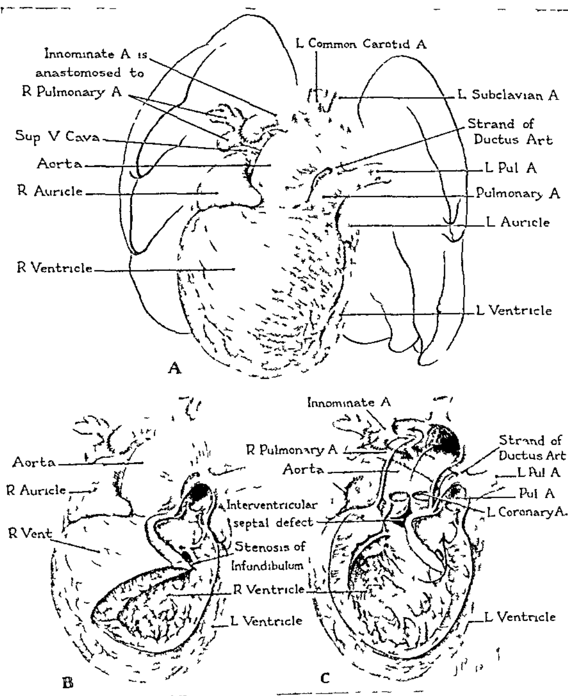


FIGURE VI-1 Tetralogy of Fallot with severe pulmonary stenosis (same patient as in Figure VI-14) Infant

The innominate artery is anastomosed to the right pulmonary artery Note the stenosis of the infundibulum and the small size of the pulmonary artery

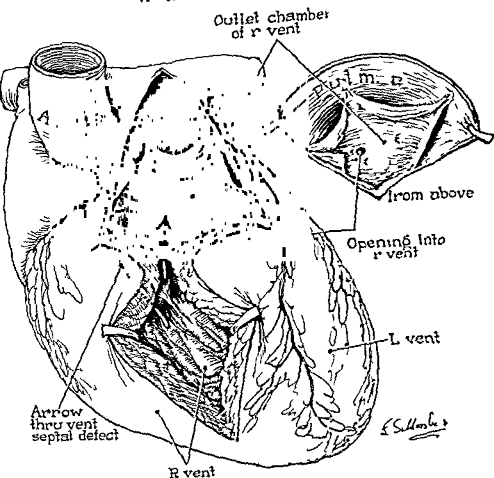


Figure vi-2 Tetralogy of Fallot with pure infundibular stenosis Adult

Note the normal pulmonary valve and the normal pulmonary artery

quently in many instances the pulmonary stenosis increases with age In all probability, patients who are able to live to adult life have less extreme degrees of pulmonary stenosis, nevertheless, it is always surprising how slender may be the thread upon which life hangs

Dextroposition of the aorta means that the aorta, although it arises mainly from the left ventricle, over rides the ventricular septum and receives some blood directly from the right ventricle The extent of the dextroposition of the aorta is subject to considerable variation Furthermore, as the aorta increases in size, it tends to over ride the ventricular septum to a greater and greater degree Occasionally the aorta may become so far dextroposed that it arises equally from both ventricles, it may even over ride the ventricular septum by more than 50 per

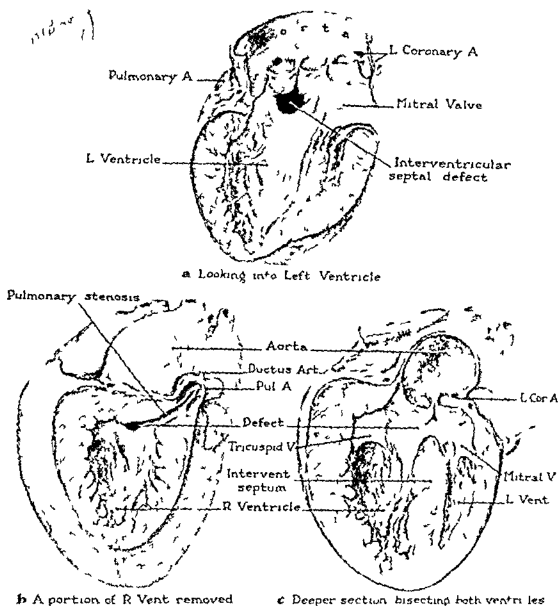


FIGURE VI-3 Tetralogy of Fallot with pulmonary stenosis Child

cent When, however, the aorta arises primarily from the right ventricle, even though it may slightly over ride the ventricular septum, the condition is classified as a transposition, not a dextroposition

Johns et al,[9] in a study of a series of patients who had died following a Blalock Taussig operation, found a high incidence of cases in which the aorta over rode the septum by more than 50 per cent This observation does not necessarily mean that such is the usual extent of the dextroposition of the aorta in the tetralogy of Fallot, because the group was composed of patients who did not survive a

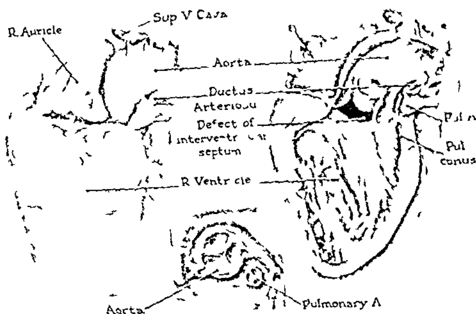


FIGURE VI-4 Tetralogy of Fallot with pulmonary atresia Infant

systemic pulmonary anastomosis One cause of death in these patients was probably the great extent of the over riding of the aorta In the classic tetralogy of Fallot, even though the pulmonary stenosis is severe, the aorta usually over rides the ventricular septum by approximately 10 per cent

A high ventricular septal defect is the inevitable result of the dextroposition of the aorta If the aorta over rides the ventricular septum, the aortic septum can not meet the ventricular septum, this causes a high ventricular septal defect The relation of the aorta to the ventricular septum is illustrated in the cross section shown in Figure VI-5 Blood does not flow from one ventricle to the other but from both ventricles into the aorta Inasmuch as some blood from the right ventricle is pumped directly into the aorta, the systemic circulation receives some venous blood there is always a venous-arterial shunt

Right ventricular hypertrophy results from the increased work required of the right ventricle This is due both to the increased volume of blood which is returned to the right ventricle and to the increased pressure against which the right ventricle must work The pulmonary stenosis and the dextroposition of the aorta both increase the work required of the right ventricle The former renders it difficult to pump the blood to the lungs the latter means that the

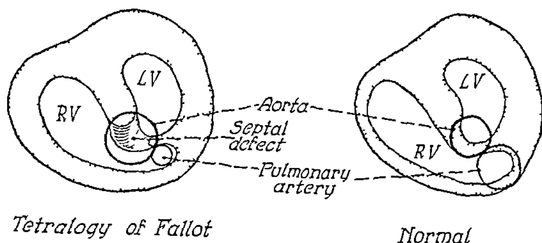


FIGURE VI-5 Cross section of a tetralogy of Fallot showing the relation of the aorta to the ventricular septum and cross section of a normal heart

right ventricle must pump against systemic pressure to eject blood into the aorta There results right ventricular hypertrophy

COURSE OF THE CIRCULATION

During fetal life, because of the dextroposition of the aorta, some blood from the right ventricle passes directly into the ascending aorta This blood mixes with the blood which flows from the pulmonary artery through the ductus arteriosus to the descending aorta (see Figure VI-6), hence a tetralogy of Fallot causes only a slight alteration in the fetal circulation At birth the heart is normal in size

After birth the foramen ovale closes promptly The ductus arteriosus, however, may remain patent for a varying period of time, as it does in all instances of diminished blood flow to the lungs Under such circumstances, owing to the low pressure in the pulmonary artery, blood will flow from the aorta through the ductus arteriosus to the lungs Moreover, because of the over riding of the aorta, the aorta receives venous as well as arterial blood, consequently, as long as the ductus arteriosus remains patent, some venous blood flows through the ductus arteriosus to the lungs The increased volume of venous blood directed to the lungs in turn increases the volume of oxygenated blood returned to the left auricle and thence to the left ventricle and to the systemic circulation In many instances the combined blood flow to the lungs through the ductus arteriosus and the stenosed pulmonary artery is sufficiently great, so that cyanosis is not visible (see Diagram VI-1)

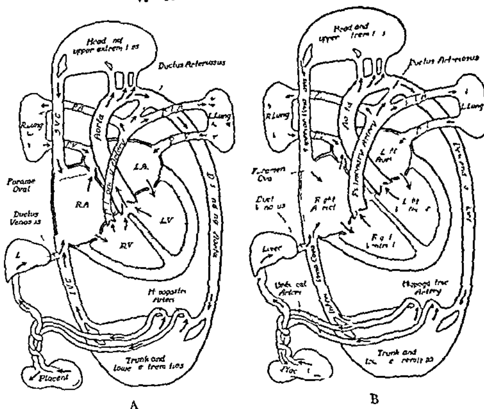
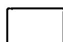


Figure vi-6 Fetal circulation (A) Tetralogy of Fallot with pulmonary stenosis and (B) normal heart

With the obliteration of the ductus arteriosus, the circulation which persists throughout life is established All the blood from the right auricle flows into the right ventricle Part of the blood from the right ventricle is pumped out by way of the stenosed pulmonary artery to the lungs and part is pumped directly into the aorta The blood which is pumped into the pulmonary artery flows to the lungs in the normal manner and the oxygenated blood is returned by the pulmonary veins to the left auricle Thence the blood flows to the left ventricle and is pumped out into the aorta The aorta not only receives all the blood from the left ventricle but, since the aorta over rides the ventricular septum, it also receives some venous blood from the right ventricle This mixture of arterial and venous blood is pumped around the systemic circulation and is returned by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again (see Diagram vi-2)

When there is pulmonary atresia the only possible way for the blood to leave the right ventricle is through the aorta, furthermore, the ductus arteriosus is

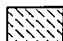
Diagram VI–1



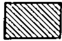
Arterial blood (fully saturated)



Venous and arterial blood
Cyanosis visible



Small admixture of venous blood
No visible cyanosis



Venous blood

Diagram VI-1

Tetralogy of Fallot with pulmonary stenosis before closure of the ductus arteriosus

The four components of the tetralogy of Fallot are (1) pulmonary stenosis, (2) dextroposition of the aorta, (3) ventricular septal defect, and (4) right ventricular hypertrophy

The dextroposition of the aorta means that the aorta over rides the ventricular septum When this occurs the aortic septum cannot meet the ventricular septum in the normal manner hence the over riding of the aorta inevitably causes a high ventricular septal defect The foramen ovale closes normally There may, however, be a delay in the closure of the ductus arteriosus When this occurs the ductus arteriosus becomes of functional importance and increases the circulation to the lungs Under such circumstances the circulation of the blood is as follows

The blood from the right auricle flows into the right ventricle Part of the blood from the right ventricle is pumped out through the stenosed pulmonary artery to the lungs and part is pumped directly into the aorta All the blood in the pulmonary arteries goes to the lungs and the oxygenated blood is returned by the pulmonary veins to the left auricle thence it passes to the left ventricle The blood from the left ventricle is pumped out into the aorta Inasmuch as the aorta over rides the ventricular septum some of the blood from the right ventricle is also pumped directly into the aorta Therefore the aorta and the systemic circulation always receive a mixture of oxygenated blood from the lungs and venous blood from the right ventricle Most of the blood in the aorta is directed to the systemic circulation and is returned by the superior vena cava and the inferior vena cava to the right auricle Owing to the pulmonary stenosis and the low pressure in the pulmonary artery, some blood from the aorta flows through the ductus arteriosus into the pulmonary artery Thus the lungs receive blood from the right ventricle and also from the aorta through the ductus arteriosus The combined blood flow to the lungs means that a relatively large volume of blood reaches the lungs All the blood which reaches the lungs is returned by the pulmonary veins to the left auricle and thence to the left ventricle Consequently a relatively large volume of oxygenated blood is pumped out from the left ventricle This blood is mixed with the relatively small volume of blood which is pumped from the right ventricle into the aorta The resultant admixture of venous and arterial blood may be insufficient to produce visible cyanosis

Clinical diagnosis The heart is normal in size and in infancy there is only a systolic murmur Therefore the clinical picture at this age closely simulates that of a ventricular septal defect The electrocardiogram in addition to the usual right axis deviation, shows evidence of right ventricular hypertrophy

Diagram VI–2



Arterial blood (fully saturated)



Small admixture of venous blood
No visible cyanosis



Venous and arterial blood
Cyanosis visible



Venous blood

DIAGRAM VI-2

Tetralogy of Fallot with pulmonary stenosis after closure of the ductus arteriosus

The four components of the tetralogy of Fallot are (1) pulmonary stenosis, (2) dextroposition of the aorta (3) ventricular septal defect, and (4) right ventricular hypertrophy

The dextroposition of the aorta means that the aorta over rides the ventricular septum When this occurs it is impossible for the aortic septum to meet the ventricular septum in the normal fashion Thus dextroposition of the aorta inevitably means that there is a high ventricular septal defect uch is the nature of the ventricular defect in the tetralogy of Fallot In this malformation the foramen ovale closes and the ductus arteriosus undergoes normal obliteration

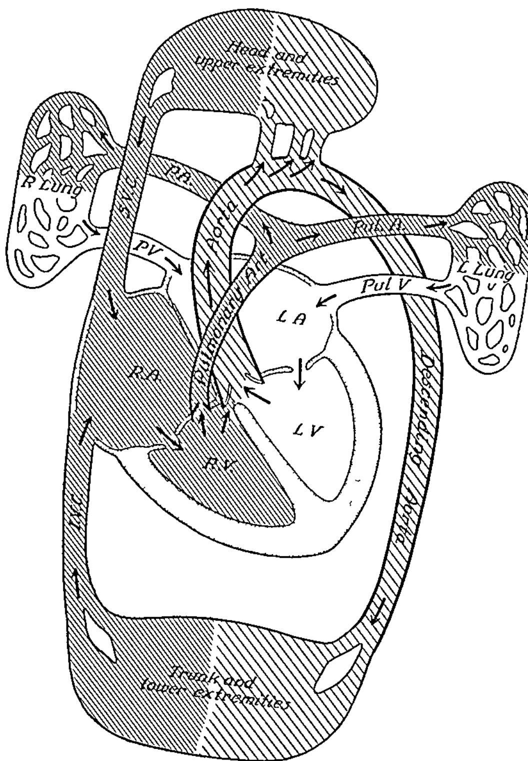
The blood which enters the right auricle flows into the right ventricle From the right ventricle part of the blood is pumped through the stenosed pulmonary orifice to the lungs where it is oxygenated and the oxygenated blood is returned by way of the pulmonary veins to the left auricle and thence passes to the left ventricle The blood from the left ventricle is pumped out through the aorta to the systemic circulation In asmuch as the aorta over rides the ventricular septum it thereby communicates directly with the right ventricle hence part of the blood from the right ventricle is directed out into the aorta Thereby the aorta and the systemic circulation receive blood from both the right ventricle and the left ventricle. All the blood in the systemic circulation is returned by the superior vena cava and the inferior vena cava to the right auricle and thence to the right ventricle Thus the right ventricle receives all the blood from the left ventri le and that part of the blood from the right ventricle which was pumped directly into the aorta. In contrast to this, the left ventricle receives only the blood from the right ventricle which was pumped through the pulmonary artery to the lungs Therefore the right ventricle always receives a greater volume of blood than does the left ventricle. Furthermore, the right ventricle has to pump the blood either through the stenosed pulmonary artery to the lungs or against systemic pressure. Hence the work of the right ventricle is increased and there results right ventricular hypertrophy

The right ventricle is enlarged and thick walled the left ventricle is small The two ventricles together give a heart of approximately normal size

The pulmonary stenosis combined with the dextroposition of the aorta means that little blood reaches the lungs for oxygenation and that a large volume of venous blood is pumped into the systemic circulation Cyanosis is usually intense

Clinical diagnosis There is usually a history of attacks of paroxysmal dyspnea in infancy and of squatting in childhood The patient shows cyanosis and clubbing of the extremities The heart is of normal size A systolic murmur is the rule. The x ray shows an absence of fullness of the pulmonary conus The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy

DIAGRAM VI–2



Arterial blood (fully saturated)

Small admixture of venous blood
No visible cyanosis

Venous and arterial blood
Cyanosis visible

Venous blood

some blood flows from the right auricle to the left auricle The greater part of the blood in the right auricle flows into the right ventricle There the cycle starts again (see Diagram VI-3)

PHYSIOLOGY OF THE MALFORMATION

In the tetralogy of Fallot, regardless of whether there is pulmonary stenosis or pulmonary atresia, only a small volume of blood reaches the lungs for oxygenation, and a large volume of blood is pumped out through the aorta to the systemic circulation It follows that the pulmonary blood flow is reduced, the systemic blood flow is increased, and the volume of blood returned to the right auricle and the right ventricle is greater than normal The right ventricle not only has an increased volume of blood to pump, but it also encounters difficulty in the expulsion of blood through the stenosed pulmonary artery Furthermore, in many instances, in order to pump blood into the aorta the right ventricle must pump against systemic pressure It follows that the ejectile force of the right ventricle must be approximately the same as that of the left ventricle Nevertheless, since blood can escape from the right ventricle into the aorta, regardless of the severity of the pulmonary stenosis, the pressure in the right ventricle seldom greatly exceeds that in the left ventricle

The right ventricle undergoes slight dilatation and considerable hypertrophy As the foramen ovale tends to close, the strain on the right auricle is increased, it, too, may become hypertrophied *The left ventricle*, however, receives only a small volume of blood and therefore remains a relatively small chamber

The pulmonary stenosis breaks the ejectile force of the right ventricle and, in addition, it reduces the volume of blood which reaches the lungs Consequently the pressure in the *pulmonary artery* is normal or abnormally low and the expansion of the lungs is normal

Inasmuch as the patency of the ductus arteriosus is not primarily determined by mechanical factors, although the closure of the ductus arteriosus is often delayed for six months or even for one year, it is usual to [illegible]

[illegible] patient with pulmonary atresia, life can be maintained only provided that there is sufficient collateral circulation to the lungs Under such circumstances the condition is functionally that of a pseudo truncus arteriosus in which the circulation to the lungs is by minute collateral vessels (see Chapter XIV) The clinical picture, however, closely resembles that of a tetralogy of Fallot with extreme pulmonary stenosis

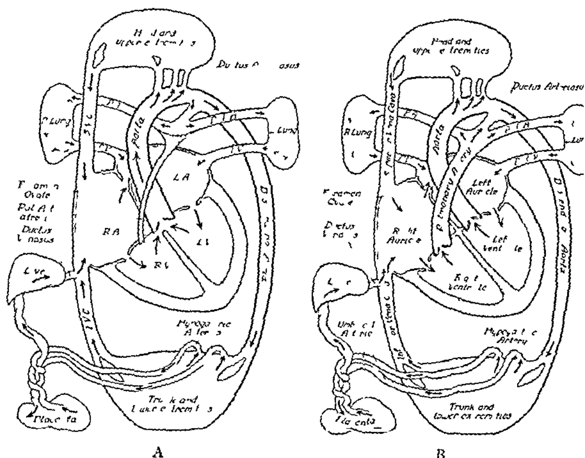


FIGURE VI-7 Fetal circulation (A) Tetralogy of Fallot with pulmonary atresia and (B) normal heart

frequently the only pathway by which the blood can reach the lungs Therefore even during fetal life blood must flow from the aorta through the ductus arteriosus into the lungs (see Figure VI-7) Since the lungs do not function and the pressure within the lungs is high, the pulmonary blood flow is very meager At birth the heart is frequently phenomenally small

Immediately after birth, with the expansion of the lungs, the pulmonary circulation is established by way of the ductus arteriosus The blood which flows from the aorta to the lungs is fully oxygenated in the lungs and is returned in the normal manner to the left auricle and thence to the left ventricle and the aorta All the blood in the aorta which is directed to the systemic circulation is returned by the superior vena cava and the inferior vena cava to the right auricle Consequently the volume of blood returned to the right auricle is greater than that returned to the left auricle and the pressure in the right auricle is higher than that in the left auricle The increased pressure in the right auricle tends to hold the foramen ovale open and therefore during the first days of life

Diagram VI-3

Tetralogy of Fallot with pulmonary atresia

When a tetralogy of Fallot is associated with pulmonary atresia and not pulmonary stenosis the course of the circulation is altered because no blood can flow through the atretic pulmonary artery to the lungs consequently the entire circulation to the lungs is by way of the ductus arteriosus Therefore unless extensive collateral circulation develops early the condition is compatible with life only as long as the ductus arteriosus remains patent.

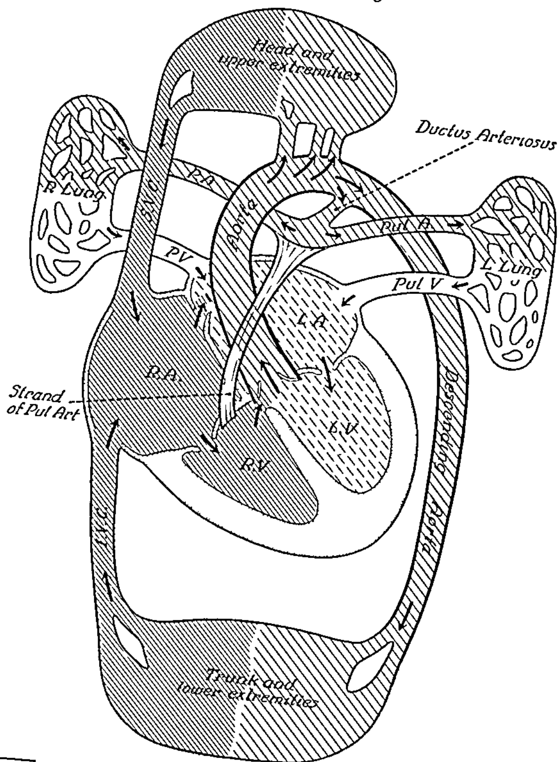
Most of the blood from the right auricle flows into the right ventricle inasmuch as there is pulmonary atresia the blood cannot be pumped from the right ventricle into the pulmonary artery All the blood from the right ventricle is pumped out into the aorta, which over rides the ventricular septum and opens directly into the right ventricle. From the aorta the greater part of the blood flows into the systemic circulation and is returned by the superior and *inferior* venae cavae to the right auricle *Inasmuch as there is* pulmonary atresia the pressure in the pulmonary artery is abnormally low and *some* blood from the aorta flows through the ductus arteriosus into the pulmonary arteries and thence to the lungs The oxygenated blood *from the lungs is returned by* way of the pulmonary veins to the left auricle Thence *it passes to the left* ventricle and is pumped out into the aorta where it mixes with the *venous blood from* the right ventricle. There the cycle starts again

The admixture of arterial and venous blood in the aorta combined with the relatively small volume of blood which reaches the lungs *for oxygenation* causes persistent cyanosis As the ductus arteriosus closes off less and less blood reaches the lungs, more and more blood is pumped through the systemic circulation Consequently the volume of blood which is returned to the right auricle becomes larger and the volume of blood to the left auricle becomes smaller As the pressure in the right auricle rises and the pressure in the left auricle falls, the valve *covering the* foramen *ovale is* forced open and the venou blood from the right auricle flows into the left auricle, thence *it* flows to the left ventricle and out into the aorta

Cyanosis becomes progressively more intense The obliteration of the ductus arteriosus usually *renders the condition* incompatible with life The duration of life and the amount of hypertrophy *of the right ventricle* depend upon the rate of obliteration of the ductus arteriosus

Clinical diagnosis Cyanosis is intense The onset of paroxysmal dyspnea [illegible] early in life The attacks may [illegible]
The infant gains weight slow [illegible]
mur if present is systolic in [illegible] Diagnosis is based primarily on x ray findings Viewed in the anterior posterior position the heart is essentially *normal* in contour with no fullness *of the* pulmonary conus In the left anterior-oblique position the right ventricle is enlarged the left ventricle is relatively small and the pulmonary window is unduly clear, the heart is like *a little* round *apple* The electrocardiogram shows a right axis deviation and evidence *of* right ventricular hypertrophy

DIAGRAM VI–3



Arterial blood (fully saturated)

Small admixture of venous blood
No visible cyanosis

Venous and arterial blood
Cyanosis visible

Venous blood

Cyanosis usually becomes evident between three and six months of age Often at first the infant suffers from attacks of cyanosis and paroxysmal dyspnea Then after a number of such episodes cyanosis becomes persistent Sometimes it is not until the infant begins to walk that cyanosis becomes apparent In rare instances, even in childhood, cyanosis is apparent only upon exercise Although the development of cyanosis is always distressing to the parents, the later its appearance, the less severe is the pulmonary stenosis Therefore, when cyanosis is not suspected until early childhood, parents can be given the consolation that the prognosis is relatively better than if the cyanosis had developed at an earlier age

The intensity of the cyanosis varies with the amount of available hemoglobin and the percentage of reduced hemoglobin in the circulating blood The latter depends upon the degree of the pulmonary stenosis and the extent of the dextroposition of the aorta After the closure of the ductus arteriosus, cyanosis is almost always present If the stenosis is extreme, cyanosis appears early and becomes intense Some patients show a purple glow over the entire body Nevertheless, it is important to emphasize that many infants may suffer from severe degrees of anoxemia with only minimal cyanosis

Polycythemia although seldom present in early infancy, is the rule in children and adults who suffer from anoxemia It usually develops simultaneously with the development of the collateral circulation, and hence after the closure of the ductus arteriosus The rate at which polycythemia develops depends upon the degree of oxygen unsaturation of the arterial blood and the demands of the body for oxygen The red blood cell count, the amount of available hemoglobin, and the hematocrit reading usually remain within normal limits for the first six months of life It is for this reason that cyanosis is so much less conspicuous in infants than in children Indeed, only in patients with severe pulmonary stenosis is there marked polycythemia by two years of age

In children and young adults the development of polycythemia is the rule The red blood cell count is usually between 6 and 10 million and may occasionally reach 11 or even 12 million The amount of available hemoglobin and the hematocrit reading are proportionally increased The hemoglobin may reach 22 grams or 140 per cent The hematocrit reading is usually between 60 and 80 per cent and in a few instances may rise above 90 per cent

The blood platelets frequently become markedly reduced and the *blood fibrinogen* becomes abnormally low, as the blood becomes abnormally thick Although the clotting time remains normal, the clot becomes extremely fragile[10] These changes probably express an attempt on the body s part to compensate for

CLINICAL FINDINGS

The clinical findings vary with the severity of the pulmonary stenosis As previously mentioned, so long as the ductus arteriosus remains open the combined flow of blood to the lungs through the pulmonary artery and through the ductus arteriosus may be *sufficient to prevent "visible" cyanosis*

It is worthy of note that in Sandifort's case and in the first case reported by Peacock both physicians stressed the fact that the patient was perfectly normal at birth Furthermore, Sandifort postulated that the normality of color during early infancy was probably due to the flow of blood through the ductus Botalli[1]

Difficulty in feeding, failure to gain, and *stunting of growth* may be the first indications of an abnormality These difficulties result from an inadequate supply of oxygen The infant gains weight very slowly because he simply cannot digest much food Small and frequent feedings may help Many infants develop severe anemia, which adds to the difficulty *Cessation of weight gain* is a serious manifestation and is usually indicative of an extremely low arterial oxygen saturation During childhood the patient generally improves but growth is slow The onset of puberty may be delayed, nevertheless, the majority of individuals with a tetralogy of Fallot *attain full physical development*

Attacks of paroxysmal dyspnea are common in infants with a tetralogy of Fallot These attacks may be precipitated by nursing or by bowel movements, occasionally they occur without provocation The infant becomes very cyanotic, respirations are rapid, and there is often difficulty in expiration These attacks may progress to loss of consciousness and may even cause convulsions They apparently occur at the time when the ductus arteriosus is undergoing obliteration The period of greatest difficulty is usually between the ages of six and eighteen months From eighteen months to two years of age the infant holds his own During this time the attacks of paroxysmal dyspnea decrease and then cease, thereafter the child starts to improve For the treatment of paroxysmal dyspnea, see below and also Chapter v

Loss of consciousness is always a serious manifestation Infants seldom lose consciousness until the oxygen saturation of the arterial blood falls to 12 per cent It may fall even lower An extremely low arterial oxygen saturation should never be dismissed as a technical error, nor should it be assumed that the sample was taken from a vein, because the oxygen saturation of the arterial blood can be extremely low When the arterial saturation is only 10 or 12 per cent, there is real danger that a further fall may be fatal Indeed, such is the probable cause of sudden death, which occasionally occurs



FIGURE VI-8 Squatting

Although the habit of squatting is characteristic of children with a tetralogy of Fallot, it is not limited to this malformation Children with a complete transposition of the great vessels combined with pulmonary stenosis also squat when tired The reason why these children squat is not known One child, who was ten years of age, replied to the author s query as to why he squatted Can t you see? It cuts off the circulation to my legs and increases the circulation to my

the increased viscosity of the blood, they lessen the danger of thrombosis but increase the patient's tendency to hemorrhage

Telangiectases and *purpuric eruptions* frequently develop over the lower extremities in patients with long standing polycythemia Such eruptions are generally associated with dilatation of the capillaries and may be related to the changes in the clotting mechanism of the blood

Clubbing of the extremities develops as the red blood cell count rises It is the result of the compensatory polycythemia, the dilatation of the capillaries, and the persistent oxygen unsaturation of the arterial blood With extreme polycythemia the fingers and toes may show drumstick deformities After successful operation even pronounced clubbing may eventually disappear

Dyspnea on exertion is common The patient's ability to exercise may be extremely limited Although the heart rate increases with exercise, owing to the pulmonary stenosis the circulation to the lungs cannot be significantly increased The consequence is that, with an increase in the minute volume, the amount of blood which reaches the lungs remains constant and the increase in the heart rate causes more venous blood to be pumped into the systemic circulation There is a fall in the oxygen saturation of the arterial blood For a short period of time the increase in the heart rate may sufficiently compensate for the fall in the arterial oxygen saturation to maintain or even to increase the supply of oxygen to the body Nevertheless, the oxygen saturation of the arterial blood falls rapidly and to such a low level that the patient develops dyspnea and a corresponding increase in the intensity of the cyanosis

Squatting is a common habit among children with a tetralogy of Fallot (see Figure vi–8) These children are able to get their breath more easily in this position Sometimes the child runs for a short distance and then suddenly squats down The author had one patient who walked in a squatting position The benefit which may be derived by squatting was delightfully illustrated by another patient, who could play football but could not participate in any other sport Football was possible because, as he said, All the children ran and squatted down and ran again '

Children with severe pulmonary stenosis frequently desire to sit crouched on their legs They usually like to cross their knees and, if allowed, will double up the knees and sit leaning slightly forward with the chest upon the knees Most of these infants and children, when extremely fatigued, automatically assume the knee chest position, lying with the weight of the thorax on the shoulders and neck



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able to work eight hours a day and complained only of slight fatigue at nightfall even though he had an oxygen saturation of the arterial blood of only 54 per cent Indeed, he was so well adjusted that he desired operation only provided it would fit him for military service Inasmuch as this was not possible, operation was not undertaken

The blood pressure is often difficult to obtain because the pulse is weak and the pulse pressure is narrow The strength of the pulse, however, is equal in arm and leg

Systemic hypertension is relatively common in patients with a tetralogy of Fallot, especially when the pulmonary stenosis is extreme The etiology is not clear It may be secondary to the renal complications which frequently occur in patients with long standing polycythemia Elevation of the nonprotein nitrogen is relatively common Furthermore, it may return to a normal level after a successful anastomosis and the blood pressure may also become lower

Hemiatrophy is relatively common in patients with a tetralogy of Fallot When it does occur, the aorta almost invariably, if not invariably, arches toward the normal side of the body This phenomenon occurs so regularly that if a patient has a *left* hemiatrophy one can postulate that he has a *right aortic arch*

CARDIAC FINDINGS

The heart is usually strikingly small The malformation in itself places a constant load on the heart and does not cause progressive cardiac enlargement Therefore the heart usually remains within normal limits Indeed, *in patients* with a severe degree of pulmonary stenosis or atresia, *the heart may be exceptionally small* The right ventricular wall is, however, always increased in thickness and may become sufficiently enlarged to press against the anterior chest wall and cause left sided chest deformity

Murmurs are frequently absent during the first days of life and the infant is considered normal By the end of the first week, *however, a* murmur is usually audible It is *systolic* in time and of maximal intensity *along the left* sternal border It may be heard posteriorly but is usually not transmitted to the vessels of the neck Such murmurs are often better heard in the *recumbent* than in the erect position but occasionally may be heard only when the patient is leaning forward

The intensity of the murmur varies *inversely with the* severity of the pulmonary stenosis A mild *pulmonary* stenosis causes a loud systolic murmur, whereas if there is extremely *severe pulmonary stenosis or pulmonary* atresia, even though there is dextroposition of the aorta, no murmur is *audible* Conse

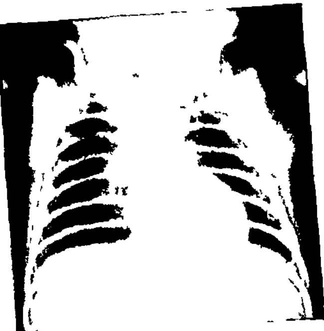
lungs" That may be the correct explanation Certain it is that these children get their breath more easily in a squatting position

The exercise tolerance of these children varies with the severity of the pul monary stenosis and with the extent of the collateral circulation It also varies in the same individual from day to day Almost every child will say that 'on good days" he can do thus and so, and that 'on bad days" he can walk only a short distance or just a few steps Indeed, on very bad days he may be bedridden Extremes of heat and cold and damp weather lessen his exercise tolerance, dry air is a great boon Consequently the dry air at a high altitude may be better tolerated than the damp air at sea level

In some children with a tetralogy of Fallot exercise causes the oxygen satura tion of the arterial blood to drop so rapidly that they can walk only a few steps before squatting to rest, whereupon the oxygen saturation of the arterial blood rises with equal rapidity and to such a height that there is no stimulus for the development of polycythemia Such patients have a normal red blood cell count and at rest may show no cyanosis Indeed, the color may be so normal that the family does not realize that the child is cyanotic The author knows of one in stance in which the parents sought the aid of a psychiatrist because the child de veloped the habit of squatting Except for the absence of cyanosis, such children have a typical tetralogy of Fallot, hence the condition has been called an "acya notic tetralogy of Fallot

The development of collateral circulation and that of polycythemia appear to go hand in hand As these increase, the oxygen saturation of the arterial blood rises The first indication of this may be a decrease in the number of attacks of paroxysmal dyspnea Later, as the child grows, his exercise tolerance usually increases He looks more cyanotic but he is stronger By the time the child reaches puberty he may be able to walk for considerable distances at a slow pace and generally no longer feels the need to squat This is due in part to the increase in the oxygen saturation of the arterial blood and in part to the fact that the collateral circulation develops from the systemic circulation Consequently an increase in the systemic blood flow tends to increase the flow of blood through the collateral vessels This results in an increase in the pulmonary blood flow

Over a period of years the oxygen saturation of the arterial blood gradually rises, so that most young adults have an oxygen saturation between 70 and 80 per cent at rest and show a less marked fall in the oxygen saturation upon exer cise Even when the oxygen saturation of the arterial blood remains low it is re markable how well these patients adjust The author had one patient who was



Anterior posterior position



Left anterior-oblique position

FIGURE VI-9 Tetralogy of Fallot with functional pulmonary atresia Infant

quently the intensity of the murmur offers a clue to the severity of the pulmonary stenosis

A systolic thrill is generally palpable if the murmur is moderately loud

The pulmonic second sound is usually faint but audible The small size of the pulmonary artery, combined with the small volume of blood which is forced through its orifice, renders the closure of the pulmonic valve difficult to hear Nevertheless, the aorta may be displaced so far to the left that the closure of the aortic valve is better heard to the left than to the right of the sternum Consequently, when the second sound to the left of the sternum is greater than the second sound to the right of the sternum, it is strong presumptive evidence of extreme dextroposition of the aorta

A third heart sound, or even a *gallop rhythm,* is seldom heard in patients with a tetralogy of Fallot The occurrence of a gallop rhythm is always indicative of a failing heart

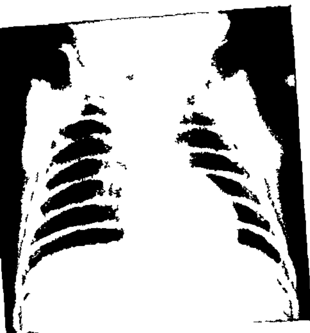
Cardiac failure is rare in patients with a tetralogy of Fallot Occasionally an infant with severe anemia and a tetralogy of Fallot may suffer from cardiac dilatation and cardiac failure Such infants develop engorgement of the liver but, owing to the inadequate blood flow to the lungs, these patients seldom suffer from pulmonary congestion

X RAY AND FLUOROSCOPIC FINDINGS

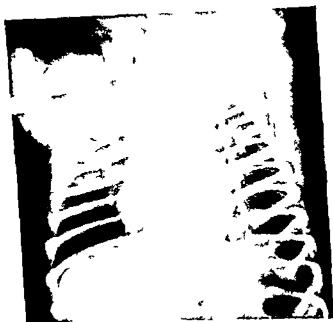
X ray and fluoroscopic findings vary with the age of the patient and the severity of the pulmonary stenosis

In infants with anatomical or functional pulmonary atresia the heart is frequently phenomenally small, as shown in Figure VI–9 (A–P position) and Figures VI–10 and 11 In the left anterior oblique position the enlargement of the right ventricle is usually apparent The heart has the appearance of a little round apple with a stem at the base This shape is caused by the enlargement of the right ventricle combined with a small left ventricle The absence of the pulmonary artery renders the pulmonary window abnormally clear and makes the aorta look like the stem of an apple, as shown in Figures VI–9, 12, and 13 (LAO position) When there is pulmonary stenosis, the heart is remarkably normal in size and shape (see Figure VI–14)

As the patient grows and the diaphragm descends, the heart assumes a characteristic contour In the anterior posterior position the heart appears to be normal in size but, owing to the stenosis of the infundibulum of the right ventricle, there is no fullness in the region of the pulmonary conus The upper margin



Anterior posterior position



Left anterior-oblique position

Figure VI-9 Tetralogy of Fallot with functional pulmonary atresia Infant



FIGURE VI-10 Tetralogy of Fallot with functional pulmonary atresia (same patient as in Figure VI-12) Infant

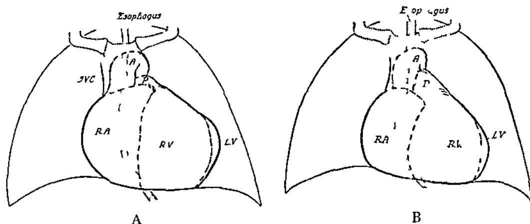


FIGURE VI-11 (A) Tetralogy of Fallot with *functional pulmonary* atresia and (B) normal heart Infant



Left anterior oblique position



Right anterior-oblique position

FIGURE VI-12 Tetralogy of Fallot with functional pulmonary atresia (same patient as in Figure VI-10) Infant



Figure VI-10 Tetralogy of Fallot with functional pulmonary atresia (same patient as in Figure VI-12) Infant

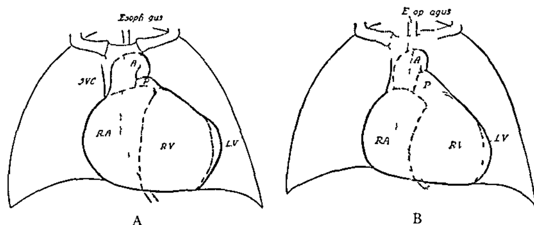


Figure VI-11 (A) Tetralogy of Fallot with functional pulmonary atresia and (B) normal heart Infant

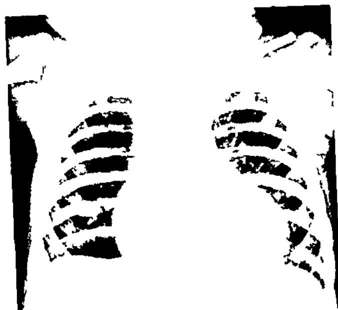
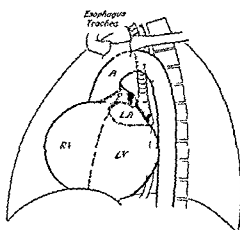


FIGURE VI-14 Tetralogy of Fallot with severe pulmonary stenosis (same patient as in Figure VI-1) Infant

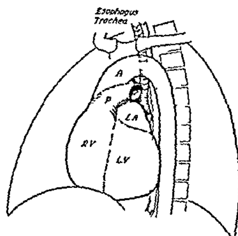
of the cardiac silhouette to the left of the sternum, in contrast to the normal contour, is concave (see Figures VI-15 and 16)

The small size of the pulmonary artery combined with the partial rotation of the great vessels causes the shadow cast by the great vessels to be narrow Consequently the contour of the heart is that of a *coeur en sabot* or 'boot shaped heart , this has been repeatedly emphasized by numerous observers[11 17] and is shown in Figures VI-17 and 18

A right aortic arch is relatively common in this malformation This condition is not to be confused with dextroposition of the aorta In the latter the aorta over rides the right ventricle, whereas the characteristic of a right aortic arch is that the aorta arches to the right Inasmuch as this variant occurs in approximately 25 per cent of all patients with a tetralogy of Fallot, the possibility should always be investigated (see Chapter XXVI) When the aorta arches to the right, the heart is usually long and narrow and its left border shows a slight concavity or a gentle slope (see Figures VI-19 and 20) The aortic knob is frequently visible to the right of the sternum within the shadow of the superior vena cava Even when the x ray or fluoroscopic examination does not show the aorta on the right, upon delineation of the esophagus by barium-opaque mixture, the esophagus is seen to lie to the left of the great vessels Under such circumstances,

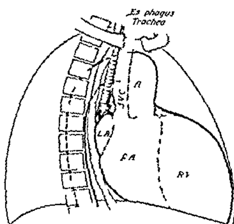


A

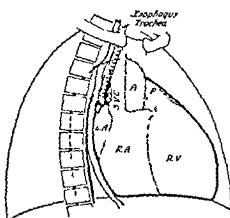


B

LEFT ANTERIOR OBLIQUE POSITION



A



B

RIGHT ANTERIOR-OBLIQUE POSITION

FIGURE VI-13 (A) Tetralogy of Fallot with functional pulmonary atresia and (B) normal heart Infant

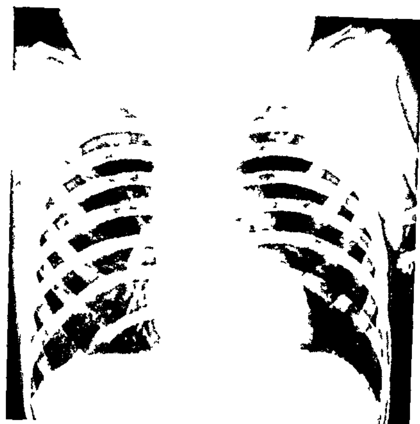


FIGURE VI-14 Tetralogy of Fallot with severe pulmonary stenosis (same patient as in Figure VI-1) Infant

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Before anastomosis

After anastomosis

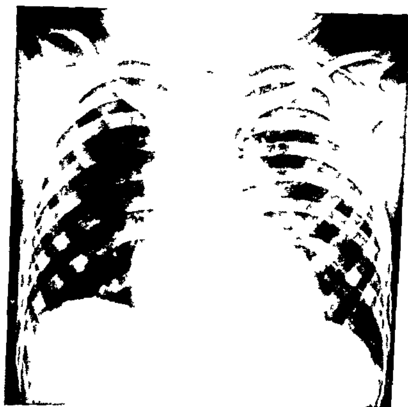
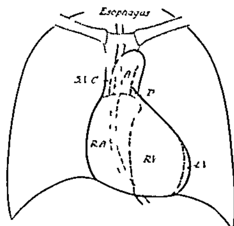
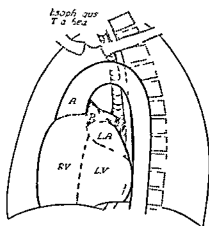


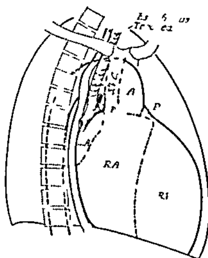
FIGURE 11-15 Tetralogy of Fallot with a right aortic arch Child



Anterior posterior position



Left anterior-oblique position



Right anterior-oblique position

FIGURE VI-16 Tetralogy of Fallot Child



Figure VI-17 Tetralogy of Fallot with a left aortic arch Adult

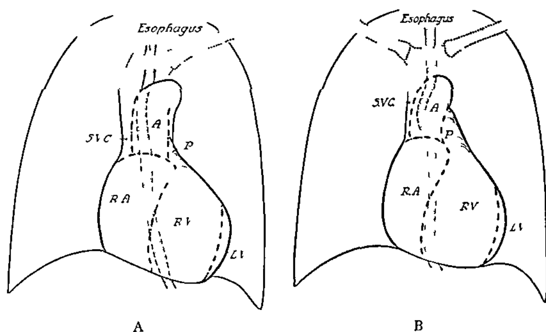


Figure VI-18 (A) Tetralogy of Fallot with a left aortic arch and (B) normal heart Adult

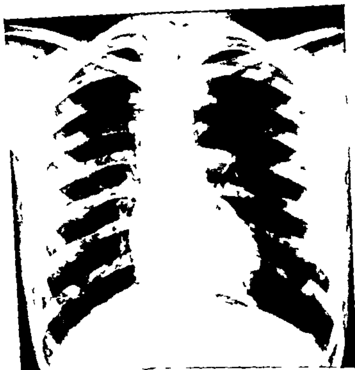


FIGURE VI-19 Tetralogy of Fallot with a right aortic arch Adult

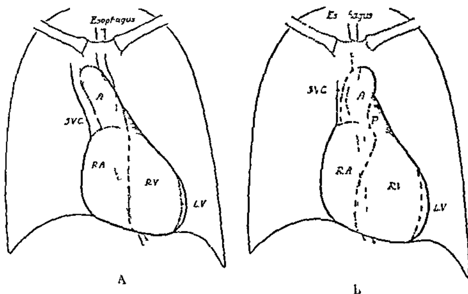


FIGURE VI-20 (A) Tetralogy of Fallot with a right aortic arch and (B) normal heart Adult

if encroached upon by the aorta, the esophagus is deviated to the left and indented on its right margin by the aorta, as shown in Figures VI-19 and 20 (see also Chapter XXVI)

In the left anterior oblique position the right ventricle is seen to be enlarged, it projects forward toward the anterior chest wall Because of the anterior displacement of the aorta, the aortic shadow lies farther from the spinal column and closer to the anterior chest wall than in the normal heart It requires minimal rotation of the patient for the left ventricle to clear the spinal column The pulmonary window is abnormally clear Furthermore, in cases of a right aortic arch, the relation of the esophagus to the aorta offers additional evidence of the existence of this anomaly *It is in this position, and not in the right anterior* oblique position, that the aorta may be seen to indent the esophagus (see Figure VI-21, LAO position, and also Chapter XXVI)

In the right anterior-oblique position there is no evidence of enlargement of the left auricle Usually there is a striking absence of the fullness of the pulmonary conus (see Figures VI-12 and 13, RAO position) Furthermore, if there is a right aortic arch, the esophagus is seen to descend in a perfectly straight line unrelated to the arch of the aorta (see Figure VI-21, RAO position)

The hilar shadows should be studied with care Owing to the diminished pulmonary circulation, the lungs are usually abnormally clear In older children a tremendous network of small vessels of collateral circulation may develop in the hili of the lungs, these may become so extensive as to cause an increase in the hilar markings If the observer s eyes are fully compensated, these shadows can be seen to be composed of small elements of varying opacity in which ex*pansile pulsations are virtually never visible*

In the evaluation of the hilar shadows it is important to remember that development of collateral circulation and that of polycythemia go hand in hand Therefore, if polycythemia *is slight or absent, increased vascular markings* should never be attributed to collateral circulation

In patients with only slight cyanosis the pulmonary artery is usually relatively large and may occasionally show slight pulsations If, however, the patient is intensely cyanotic, there should be no pulsations in these shadows Furthermore, as the child grows, the heart and great vessels increase in size It follows that the pulmonary artery is more readily visible in older children than in infants Consequently the hilar shadows should always be evaluated in relation to the patient s age and the clinical findings

Slight fullness of the pulmonary conus occurs in a small percentage of pa

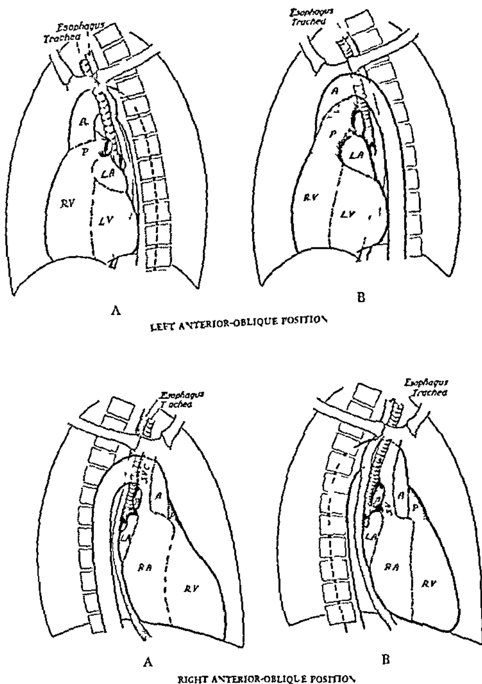


FIGURE VI-21 (A) Tetralogy of Fallot with a right aortic arch and (B) normal heart Adult

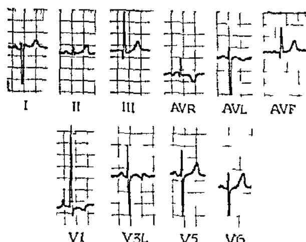


FIGURE VI-22 Tetralogy of Fallot

tients In such instances the pulmonary arteries are readily seen and appear to be of normal size This contour is more frequently encountered in young adults than in infants, partly because the vessels are larger and partly because the pulmonary stenosis is relatively mild Indeed, many of these individuals do so well that medical attention is not sought in infancy and early childhood When the pulmonary artery is relatively large, it raises the question of valvular pulmonary stenosis combined with a fair sized infundibular chamber, or of a valvular pulmonary stenosis with an intact ventricular septum Patients who show such features may require cardiac catheterization, angiocardiography, or both to confirm the diagnosis

ELECTROCARDIOGRAPHIC FINDINGS

A definite right axis deviation in the standard leads and evidence of right ventricular hypertrophy in the unipolar precordial leads are characteristic of this malformation, but there is rarely the prolongation of the intrinsicoid deflection or the deep inversion of T waves in V_1, such as is commonly seen in severe pulmonary stenosis with an intact ventricular septum Absence of right ventricular hypertrophy in the precordial leads is a reason to doubt the diagnosis and is a definite indication for further studies The P waves are frequently abnormally high and pointed (see Figure VI-22) The T waves may or may not be abnormal

SPECIAL TESTS

The circulation time (venous to systemic) is abnormally short In children it is usually between four and six seconds and in adults between eight and twelve seconds The occurrence of a twenty second circulation time should always arouse suspicion that the aorta does not over ride the right ventricle

The oxygen saturation of the arterial blood is abnormally low and falls still lower with exercise A rapid fall in the oxygen saturation of the arterial blood with exercise is characteristic of this malformation When there is doubt as to the presence of cyanosis, exercise will usually render the cyanosis readily apparent

The author had one patient in whom the oxygen saturation of the arterial blood dropped from 83 per cent to 33 per cent on slight exertion, and another in whom it fell from 41 per cent to 15 per cent upon climbing two steps three times When the oxygen saturation of the arterial blood is between 30 per cent and 35 per cent the child can walk only a few feet When the oxygen saturation is between 20 per cent and 25 per cent the child is seldom able to walk Such a child has barely enough oxygen to meet the basal requirements of the body

During infancy the oxygen saturation of the arterial blood may be extraordinarily low It is quite common to find that the arterial oxygen saturation drops to 30 per cent with exercise or vigorous crying If there is a harsh systolic murmur, such a drop in oxygen saturation need cause no undue concern If the oxygen saturation drops to 20 per cent or less, there is reason for concern Most infants lose consciousness when the oxygen saturation drops to 10 per cent or less For such an infant early operation is indicated, especially if the murmur is faint or absent

Effect of exercise on oxygen consumption Usually the pulmonary stenosis is so severe that the patient is unable appreciably to increase the pulmonary blood flow with exercise Consequently exercise causes both a fall in the oxygen saturation and a fall in the oxygen consumption per liter of ventilation[18] This test is however, not diagnostic of a tetralogy of Fallot

Cardiac catheterization may be of aid in the confirmation of the diagnosis, especially when there is fullness of the pulmonary conus or there are slight pulsations in the pulmonary arteries In order to prove the existence of pulmonary stenosis it is essential to catheterize the pulmonary artery and to obtain pressure readings or pressure tracings in the pulmonary artery and in the right ventricle The finding of high pressure in the right ventricle, combined with low pressure in the pulmonary artery, is indicative of pulmonary stenosis In a tetralogy of Fallot, due to the over riding of the aorta, the pressure in the right ventricle usually approaches but seldom exceeds the systemic pressure The author has, however, seen one patient with a tetralogy of Fallot and an extremely small pulmonary artery who ten years after a successful Blalock Taussig operation had developed extreme right ventricular hypertrophy and had a pressure of 250 mm

of mercury in his right ventricle If the catheter enters the aorta, it usually means that the aorta arises in part or entirely from the right ventricle

Although the shunt is primarily from right to left, there is usually a small left to right shunt Therefore it is *common to find a slight increase in the oxygen* content of the sample of blood taken from the right ventricle as compared with that taken from the right auricle, especially when the sample is taken from the outflow tract of the right ventricle, frequently the oxygen content of the blood in the pulmonary artery is slightly higher than that in the right ventricle This finding is consistent with, but not diagnostic of, a tetralogy of Fallot

Cardiac catheterization can definitely establish the diagnosis of a tetralogy of Fallot, provided that both the aorta and the pulmonary artery are catheterized and that pressure tracings and blood samples are obtained from the various chambers This is, however, seldom possible Furthermore, catheterization gives no clear indication of the degree of dextroposition of the aorta Inasmuch as cardiac catheterization seldom definitively establishes the diagnosis, if the clinical findings are characteristic of a tetralogy of Fallot catheterization is not a prerequisite for operation

*Angiocardiography is the most useful test in the demonstration of dextro*position of the aorta When the aorta arises in part or entirely from the right ventricle, it is the rule to see dense filling of the aorta which occurs simultaneously with the filling of the pulmonary artery (see Figure VI-23) This, however, does not always occur The author has studied one infant in whom, although the aorta did not fill until after the dye had circulated through the lungs and returned to the left auricle and left ventricle, operation revealed a fairly large ventricular septal defect and an over riding aorta Furthermore, angiocardiography does not give reliable evidence concerning the size of the pulmonary artery The pulmonary artery may appear to be of moderate size in the angiocardiogram and prove to be extremely small at operation, or the reverse may be true

The aorta is usually better visualized in the anterior posterior view than in the lateral view Therefore, when only a small amount of dye is shunted into the aorta, early delineation of the aorta may be seen in the anterior posterior film and not in the lateral film Nevertheless, lateral films are essential in order to be sure that there is no dye in the left auricle If dye is immediately shunted into the left auricle, there is *either an auricular defect or patency* of the foramen ovale In either case dye may reach the aorta by way of the left ventricle simultaneously with the filling of the pulmonary artery from the right ventricle Con

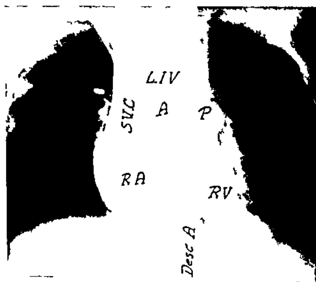




FIGURE VI–23 Tetralogy of Fallot Child

of mercury in his right ventricle If the catheter enters the aorta, it usually means *that the aorta arises in part or entirely from the right ventricle*

Although the shunt is primarily from right to left, there is usually a small left-to-right shunt Therefore it is common to find a *slight increase in the oxygen* content of the sample of blood taken from the right ventricle as compared with that taken from the right auricle, especially when the sample is taken from the outflow tract of the right ventricle, frequently the oxygen content of the blood in the pulmonary artery is slightly higher than that in the right ventricle This finding is consistent with, but not diagnostic of, a tetralogy of Fallot

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trophy As the ductus arteriosus undergoes obliteration, the infant with a tetralogy of Fallot usually develops paroxysmal dyspnea and persistent cyanosis

In an older patient the history is significant in that cyanosis usually dates from infancy and that in childhood squatting was a common habit Physical examination reveals cyanosis and clubbing, in combination with a heart of normal size Usually there is a systolic murmur and a thrill maximal along the left sternal border The second sound at the base is faint and pure The x ray shows a concave curve at the base of the heart to the left of the sternum The pulmonary arteries may or may not be seen The aorta may arch either to the right or to the left. Upon fluoroscopy hilar pulsations are minimal or absent The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy

In general, the more severe the pulmonary stenosis, the clearer are the lung fields and the fainter is the murmur In older patients collateral circulation develops and polycythemia increases With intense cyanosis and marked polycythemia, the hilar shadows may become accentuated If however, there is no polycythemia, dense hilar shadows should not be attributed to collateral circulation By and large it is rare for an adult with a tetralogy of Fallot to have *no murmur* because it is rare for an individual with a tetralogy of Fallot and a severe pulmonary stenosis to live to adult life If there is no murmur and also no great limitation of activity, the patient does *not* have a tetralogy of Fallot

DIFFERENTIAL DIAGNOSIS

A tetralogy of Fallot must be differentiated from a small ventricular septal defect tricuspid atresia, truncus arteriosus with reduced pulmonary blood flow, and complete transposition of the great vessels with or without pulmonary stenosis, sometimes from a single *ventricle with* pulmonary atresia, or even from pure pulmonary stenosis, and occasionally from primary pulmonary hypertension or defective *development* of the right ventricle with pulmonary stenosis, or even from the Eisenmenger complex and Ebstein's anomaly of the tricuspid valve

A ventricular septal defect of the maladie de Roger type may require differentiation from a tetralogy of Fallot in early infancy before the appearance of cyanosis A simple ventricular septal defect may, in early infancy, be associated with a slight right axis deviation, but the precordial leads seldom show evidence of hypertrophy of either the right or the left ventricle [illegible]

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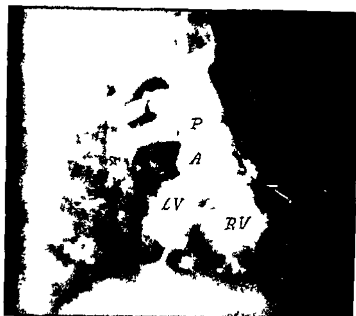


FIGURE VI-24 Tetralogy of Fallot Child

Selective angiocardiogram with dye injected into the outflow tract of the right ventricle

sequently, in the presence of a shunt at the auricular level, it is impossible to prove the dextroposition of the aorta by a *venous* angiocardiogram In such instances, selective angiocardiography with dye injected into the right ventricle eliminates the possibility of a shunt at the auricular level This technique is also best for the visualization of the outflow tract of the right ventricle, as shown in Figure VI-24

DIAGNOSIS

A tetralogy of Fallot with severe pulmonary stenosis or pulmonary atresia is to be suspected in an infant with a heart of normal size and no murmur, who has ceased to gain weight and suffers from severe attacks of paroxysmal dyspnea If the x ray shows a small heart and phenomenally clear lung fields and the electrocardiogram shows both a right axis deviation and evidence of right ventricular hypertrophy, such an infant almost certainly has a tetralogy of Fallot and is in danger of dying from anoxemia

A tetralogy of Fallot with pulmonary stenosis is to be suspected in a young infant with normal color, a heart of normal size, and a harsh systolic murmur, if the electrocardiogram shows evidence of definite right ventricular hyper

Pulmonary stenosis with an intact ventricular septum may occasionally re quire differentiation from a tetralogy of Fallot In early infancy it is the normal ity of the heart in the two conditions which causes the confusion Infants with pure pulmonary stenosis usually gain better than those with a tetralogy of Fallot and they do not suffer from attacks of paroxysmal dyspnea Evidence of progressive cardiac enlargement should always suggest that the pulmonary stenosis is associated with a closed ventricular septum The electrocardiogram frequently shows evidence of right ventricular ' strain in pure pulmonary ste nosis and only rarely does this occur in a tetralogy of Fallot

In older children and young adults the contours of the heart may occasion ally be quite similar in the two conditions A history of squatting, combined with marked limitation of exercise and a sharp fall in the oxygen saturation of the arterial blood upon exercise, is strongly suggestive of a tetralogy of Fallot

In the rare patient who, upon fluoroscopy, does show fullness of the pulmonary conus and slight pulsations in the main branches of the pulmonary artery, cardiac catheterization or angiocardiography may be necessary in order to de termine whether or not the aorta over rides the right ventricle If, upon cathe terization, the catheter enters the aorta, it is proof positive that the aorta arises in part or entirely from the right ventricle On the other hand, if the pressure in the right ventricle is definitely higher than the systemic pressure, that is usually good evidence of an intact ventricular septum Angiocardiography shows that in a tetralogy of Fallot the dye enters the aorta promptly and in a high concen tration whereas in pure pulmonary stenosis the dye lingers for a long time in the main pulmonary artery

Primary pulmonary hypertension may be confused with a tetralogy of Fallot during the first months of life Both malformations may cause deep cyanosis Furthermore, the contours of the heart may be closely similar before the pulmonary artery becomes dilated In primary pulmonary hypertension the infant, though cyanotic, does not suffer from attacks of paroxysmal dyspnea, the pul monary second sound is usually accentuated Over a period of months the heart shows progressive cardiac enlargement, which immediately differentiates the condition from a tetralogy of Fallot In doubtful cases cardiac catheterization will readily differentiate the two conditions, as in a tetralogy of Fallot the pul monary pressure is low, whereas in a primary pulmonary hypertension it is high

The Eisenmenger complex should not be confused with a tetralogy of Fallot The late development of cyanosis, the increased vascularity of the hilar mark ings, and the presence of a hilar dance all differentiate this condition from a tetralogy of Fallot In the less severe cases the murmur in a tetralogy of Fallot

to be a tetralogy of Fallot Nevertheless, as long as the infant is doing well and gaining weight there is no cause for concern and no necessity for special studies to determine the exact nature of the malformation

Tricuspid atresia is associated with electrocardiographic evidence of a left axis deviation and of left ventricular hypertrophy This finding readily differentiates it from a tetralogy of Fallot

Truncus arteriosus with markedly reduced pulmonary blood flow may be confused with a tetralogy of Fallot in which there is a severe degree of pulmonary stenosis in early infancy In a truncus arteriosus the heart is usually enlarged and occupies a horizontal position and the pulmonary artery cannot be visualized Cyanosis dates from birth and is intense, polycythemia develops early but in spite of this the infant does better than might be anticipated Such infants rarely suffer from attacks of paroxysmal dyspnea

Complete transposition of the great vessels may occasionally be confused with a tetralogy of Fallot In infants with a tetralogy of Fallot the lungs are remarkably clear; whereas in complete transposition of the great vessels the vascular markings extend nearly to the periphery of the lungs

During infancy, when a complete transposition of the great vessels is combined with a severe pulmonary stenosis, the condition may be extremely difficult to differentiate from a tetralogy of Fallot in which the pulmonary stenosis is severe The quality of the second sound at the base is of prime diagnostic value In a tetralogy of Fallot with pulmonary stenosis, the second sound at the base to the left of the sternum is faint, whereas in a complete transposition of the great vessels the aorta lies far to the left and close to the sternum and consequently the second sound in this area is accentuated

Complete transposition of the great vessels may occasionally be compatible with life for a number of years, especially when there is pulmonary stenosis In childhood this condition may be confused with that of a tetralogy of Fallot with extreme pulmonary stenosis Patients with both types of malformations are severely incapacitated and frequently squat when tired Cyanosis and polycythemia are intense In patients with complete transposition of the great vessels, stunting of growth may be extreme The systolic murmur is usually not very loud, but the second 'pulmonic' sound is loud and may be reduplicated The confusion in diagnosis arises because the increased vascular markings are interpreted as due to collateral circulation In a complete transposition of the great vessels the vascular markings extend to the periphery of the lungs, furthermore, they are usually better seen in the x ray film than upon fluoroscopy (see Chapter x)

when one pulmonary artery is well visualized and no dye enters the other pulmonary artery (see Figure VI-25) Needless to say, if an anastomosis is to be performed, it must be performed on the side on which there is a pulmonary artery

Pulmonary insufficiency occurs in the rare instances in which the pulmonary valves are absent Such cases, which are similar to the one mentioned in Chapter II have been reported by Miller et al [4] The pulmonary insufficiency causes an early diastolic murmur which is audible along the left sternal border Such a murmur may also be caused by aortic insufficiency, but this is rare and almost invariably due to acquired disease, therefore it is seldom heard in infancy

A partial anomaly of the venous return may occur Such an anomaly is compensatory in that it increases the oxygen content of the blood in the right ventricle If total correction is undertaken, this abnormality, too, will require correction

Large ventricular septal defects may be associated with mild pulmonary stenosis In such instances there is a large left to-right shunt and sufficient pulmonary stenosis to break the force with which the blood is ejected to the lungs The increased work required of the right ventricle causes the stenotic area to hypertrophy and consequently the pulmonary stenosis may become progressively greater Over a period of years some infants who suffer from a large left to-right shunt develop pulmonary stenosis Occasionally the pulmonary stenosis becomes so great that the direction of the shunt is reversed and the patient suffers from reduced pulmonary flow Thus the condition develops into a tetralogy of Fallot Gasul et al have reported a number of such patients who in early infancy suffered from a large left to-right shunt and who subsequently suffered from reduced pulmonary blood flow When closure of the ventricular septal defect becomes a safe procedure for infants, such infants may be benefited by early correction of the defect (see Chapter XXII)

COMPLICATIONS

The major complications are due to anoxemia and polycythemia and to the marked increase in the viscosity of the blood, and also to the alteration in its clotting mechanism which develops to compensate for the ...

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is always loud, whereas in an Eisenmenger complex it is subject to great variation in intensity The second sound is never reduplicated in a tetralogy of Fallot, whereas reduplication of the second sound is common in the Eisenmenger complex Furthermore, in the latter condition cardiac catheterization shows high pressure in the pulmonary artery as well as in the right ventricle

A single ventricle with reduced pulmonary blood flow may be confused with a tetralogy of Fallot Usually either the contour of the heart or the electrocardiogram suggests that the condition is not that of a tetralogy of Fallot Since the rudimentary chamber occupies the position of the outflow tract of the right ventricle, the contour becomes more nearly square than is common in a tetralogy of Fallot The electrocardiogram may show a right axis deviation, but generally does not show the usual pattern of right ventricular hypertrophy in the precordial leads Often there is a deep S wave in all six precordial leads

Defective development of the right ventricle with pulmonary stenosis and an intact ventricular septum may closely simulate a tetralogy of Fallot The absence of paroxysmal dyspnea and failure to squat differentiate this malformation from a tetralogy of Fallot Furthermore, the electrocardiogram frequently shows evidence of the transition from right to left ventricular preponderance in V_2 or V_3 (see Chapter IX, Section A)

Ebstein's anomaly of the tricuspid valve may occasionally be confused with a tetralogy of Fallot In the former condition the infant does not suffer from attacks of paroxysmal dyspnea The heart may be greatly enlarged, and the liver is often engorged but no pulsations are palpable at its margin In infants the electrocardiographic evidence of low voltage in V_1, which is so characteristic of Ebstein's anomaly, is seldom seen In older children, however, the electrocardiogram is frequently of diagnostic aid

COMMONLY ASSOCIATED MALFORMATIONS

A gross defect in the auricular septum is probably the commonest additional malformation This combination of abnormalities is sometimes spoken of as a *pentalogy of Fallot* The condition usually causes no difficulty and is detected only by cardiac catheterization Recognition of the auricular defect is, however, of importance in the surgical treatment of a tetralogy of Fallot (see below)

A single pulmonary artery occasionally occurs in association with a tetralogy of Fallot [19] Nadas et al [20] have also reported the occurrence of unilateral pulmonary atresia The difference in the vascular shadows in the x ray film offers the clue to the diagnosis This diagnosis is readily confirmed by angiocardiography

fluid intake This is especially important in patients with a tetralogy of Fallot, as the malformation is amenable to surgery and consequently compatible with relative longevity For this reason an assiduous effort should be made to prevent cerebral thrombosis and the permanent handicap which results from a hemiplegia

Brain abscess is also a very serious complication Persons in whom venous blood is shunted directly into the systemic circulation are especially prone to such an infection The liability to abscess formation is increased by the minute thrombi and consequent injury to the brain which occurs with long standing polycythemia The differentiation between a brain abscess and cerebral thrombosis is extremely important (see Chapter v) Before the age of two cerebral thromboses are common and brain abscesses are rare Generally the onset is insidious, the symptoms are progressive, and the patient runs a low grade fever If there is any possibility that a brain abscess exists, an immediate effort should be made to establish [illegible]

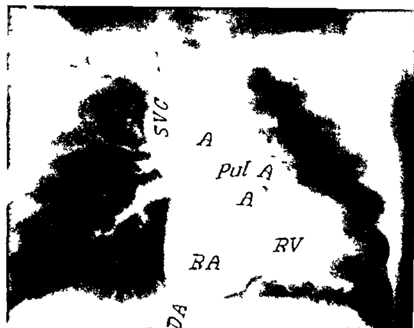
[illegible]

serious complication Therefore the patient should receive the usual preventive and therapeutic treatment (see Chapter v) If there is any suspicion of a hidden infection, operation for the amelioration or correction of a tetralogy of Fallot should never be undertaken until the danger of infection has been *eliminated* This is an extremely important precaution, because a fresh-cut surface or an anastomosis renders these patients extremely susceptible to subacute bacterial endocarditis

Fever may be caused by severe anoxemia for this reason if a patient has severe anoxemia and no demonstrable infection, operation should not be postponed because of a persistent low *grade fever*, as such a patient is in danger of dying from anoxemia

Hemoptysis is a rare complication in patients with a tetralogy of Fallot In most instances the reduction in the pulmonary blood flow is so great that hemoptyses seldom occur Nevertheless, some patients with extensive collateral circulation do develop hemoptyses, these are due in part to the great strain placed upon the small arterial vessels which lead to the lungs and in part to the bleeding tendency which develops when polycythemia becomes excessive (see Chapter iv)

Hematemesis or *intestinal bleeding* may also occur in a patient with long standing polycythemia especially when, as in a tetralogy of Fallot, the condi



Auricular diastole and ventricular systole



Auricular systole and ventricular diastole

FIGURE VI-25 Tetralogy of Fallot with a single pulmonary artery Child

and the hematocrit reading back to approximately normal values and overcomes these dangers

Surgical treatment is of great benefit to patients with a tetralogy of Fallot Inasmuch as their incapacity is primarily due to insufficient pulmonary blood flow combined with a persistent venous-arterial shunt, any operation which increases the circulation to the lungs ameliorates the condition A Blalock Taussig operation,[1] namely, the anastomosis of a systemic artery to a branch of the pulmonary artery, gives excellent results The same is true of a Potts anastomosis,[1] that is, an anastomosis between the descending aorta and the pulmonary artery It is also possible to insert a graft between the aorta and the pulmonary artery Sir Russell Brock[7] advocates the excision of the stenotic area Lillehei[8] recommends closure of the ventricular septal defect and excision of the stenotic area under direct vision Each operation has its merits and its limitations

The Blalock Taussig operation places remarkably little strain on the heart Immediately after operation cardiac enlargement occurs to adjust to the altered circulation and the increased demands placed upon the heart Thereafter there is no further cardiac enlargement Indeed, if the anastomosis fails to grow with the child, the load on the heart is lessened and it, too, fails to grow with the child, consequently the heart again becomes relatively small

The standard operation for children is the anastomosis of the proximal end of the subclavian artery which arises from the innominate artery to the side of the right or left pulmonary artery Hence the operation is performed on the opposite side to the arch of the aorta The end to-side anastomosis permits the flow of blood to both lungs After operation a good continuous murmur should be audible over both lungs posteriorly The course of the circulation after operation is shown in Diagram VI-4

The mortality rate in the last one hundred patients operated upon between two and twelve years of age was 2 per cent, hence it is a remarkably safe operation for young children

The ideal time for operation is between eight and twelve years of age Only 5 per cent of the patients in this age group have required a second operation within ten years, whereas among younger patients, that is, between four and eight years of age, 32 per cent have required a second operation within ten years but only 4 per cent have had a second operation w[illegible]

th[illegible]

[illegible] benefit (see below)

tion is associated with a large systemic blood flow and a small pulmonary blood flow.

Hemorrhagic diatheses develop in patients with long standing polycythemia. Such alteration in the blood plasma increases the risk of operation and therefore, whenever possible, operation should be performed before such changes have been of long duration. In the author's experience, even though alteration in the clotting mechanism has been demonstrated in young individuals, postoperative bleeding has not been a serious complication in patients under twelve years of age.

TREATMENT

Medical and surgical treatment are both important.

Medical treatment is mainly directed to the relief of paroxysmal dyspnea and to the prevention of complications. For a detailed discussion of the following conditions, see Chapter v.

An attack of paroxysmal dyspnea calls for prompt treatment. The infant should be immediately placed in the knee-chest position or held over the mother's shoulder with his legs doubled up beneath him. This simple procedure frequently relieves the attack. Oxygen may help to lessen the dyspnea. If the attack is long and severe or progresses to loss of consciousness, the infant should be given morphine in full doses, as its action is almost specific. The dose is 1 mgm per 4.5 kg (i.e., 10 lb.) of body weight. Within ten to fifteen minutes after receiving morphine intramuscularly the infant usually breathes more easily, his color improves, and the attack gradually wears off.

Prophylactic chemotherapy during infections and prior to and following dental extraction or tonsillectomy is extremely important both to prevent subacute bacterial endocarditis and to lessen the danger of brain abscess. Intensive chemotherapy is indicated in septicemia, in severe injuries, and also for women during delivery.

The author is not yet convinced that prophylactic antibiotics are indicated in an effort to prevent sporadic cases of *subacute bacterial endocarditis*, in part because if it is to be effective, medication must be given for life, and in part because 100 per cent protection is not humanly possible.

Adequate fluid intake in hot weather and during episodes of fever aids in the prevention of dehydration, and thereby lessens the risk of cerebral thrombosis, hemiplegia, and that rare but tragic complication—*blindness*. Successful operation brings the red blood cell count, the level of the available hemoglobin,

DIAGRAM VI-4

Tetralogy of Fallot with pulmonary stenosis after closure of the ductus arteriosus and after the Blalock Taussig operation

In a tetralogy of Fallot after the creation of an artificial ductus arteriosus by the anastomosis of the proximal end of the subclavian artery to the side of the pulmonary artery the circulation of the blood is essentially the same as it was before the closure of the ductus arteriosus

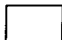
The blood from the right auricle flows into the right ventricle Part of the blood from the right ventricle is pumped out through the stenosed pulmonary artery to the lungs and part is pumped directly into the aorta All the blood in the pulmonary arteries goes to the lungs and the oxygenated blood is returned by the pulmonary veins to the left auricle and thence it passes to the left ventricle. The blood from the left ventricle is pumped out into the aorta Inasmuch as the aorta over rides the ventricular septum some of the blood from the right ventricle is also pumped directly into the aorta Therefore the aorta and the systemic circulation always receive a mixture of oxygenated blood from the lungs and venous blood from the right ventricle

Most of the blood in the aorta is directed to the systemic circulation and is returned by the superior vena cava and the inferior vena cava to the right auricle Owing to the pulmonary stenosis and the low pressure in the pulmonary artery some blood from the aorta flows through the artificial ductus arteriosus into the pulmonary artery Thus the lungs receive blood from the right ventricle and also from the aorta through the artificial ductus arteriosus The combined blood flow to the lungs means that a relatively large volume of blood reaches the lungs All the blood which reaches the lungs is returned by the pulmonary veins to the left auricle and thence to the left ventricle Consequently a relatively large volume of oxygenated blood is pumped out from the left ventricle This blood is mixed with the relatively small volume of blood which is pumped from the right ventricle into the aorta The resultant admixture of venous and arterial blood may be insufficient to produce visible cyanosis

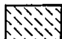
Clinical diagnosis Color is excellent The lips are of normal color The fingers may show slight cyanosis The clubbing recedes The heart may be slightly larger than before operation Its contour is essentially the same The systolic murmur, audible before operation usually becomes harsher In addition, a loud continuous murmur is heard throughout the chest this continuous murmur is usually better heard posteriorly than anteriorly

Diagram VI–4

Subclavian Artery anastomosed to Right Pul Art
Head and upper extremities
R Lung
R.A.
S.V.C.
P.V.
Aorta
Pul. A.
L Lung
Pul V
L A
Pulmonary Artery
R.A.
L V
R.V.
Descending Aorta
I.V.C.
Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

be sufficiently small to protect the lungs from the high pressure in the right ventricle which is secondary to the over riding of the aorta In the rare instance in which a tetralogy of Fallot has only a valvular pulmonary stenosis, care must be taken not to convert the malformation into an Eisenmenger complex Such an operation may be fatal, as the patient lacks the protective pulmonary hypertension characteristic of an Eisenmenger complex Indeed, unless the septal defect is closed, excision of the stenotic area is not advisable

Corrective surgery is now possible Lillehei[6] was the first to demonstrate that with the aid of a pump and oxygenator it was possible to excise the stenotic area and close the ventricular defect and thereby to restore the heart and circulation to normal Such an operation is probably the ideal for adults and for children who have nearly attained their growth

For infants and young children with severe pulmonary stenosis or pulmonary atresia, in whom the pulmonary artery is but one third to one half as large as the aorta, it seems doubtful that the right ventricle, the pulmonary orifice, and the pulmonary artery will expand to normal size and grow normally with the child When a patch is necessary, it must be made of the proper size for the patient It is manifestly impossible to insert the patch of suitable size for an adult into a tiny infant, hence there is real danger that total correction in infancy or early childhood may leave the individual with an underdeveloped right ventricle and pulmonary stenosis by the time he reaches adolescence

Furthermore, recent experience has shown that a previous Blalock anastomosis does not greatly increase the risk of operation for total correction Moreover, Ferencz [7] studies on the pulmonary vascular bed have shown that, whereas most infants with anoxemia and decreased pulmonary blood flow have multiple thrombi in their lungs, patients with an adequate anastomosis who have died of unrelated causes have a normal pulmonary vascular bed Therefore the child who has had a previous anastomosis may be better able to adjust to a corrective procedure than the patient who has not had a previous operation

The indications for operation vary with the age of the patient and the severity of the pulmonary stenosis A baby with a tetralogy of Fallot who has a small heart but no murmur, and excessively clear lung fields, may be in real danger of dying from anoxemia, even though he has no polycythemia and but slight cyanosis Such infants frequently suffer from repeated attacks of paroxysmal dyspnea If the infant ceases to gain weight and the attacks of paroxysmal dyspnea become long and severe or progress to loss of consciousness, early operation is urgently indicated If, however, the infant is gaining weight and doing well, it

In infants the problem is more difficult The subclavian artery is often too small to be of great benefit More than 33 per cent of patients operated on at the Johns Hopkins Hospital who were under two years of age have required a second operation within ten years Such infants require a relatively large vessel The sacrifice of the innominate artery has, however, proved too dangerous to the cerebral circulation to justify its use in an effort to obtain a larger vessel When a large opening is desired, a Potts procedure is preferable to a Blalock Taussig anastomosis

Potts' anastomosis, that is, an anastomosis between the descending aorta and the pulmonary artery, gives excellent results Potts has emphasized the importance of the correct size of the opening He recommends that the length of the incision be exactly 4 mm regardless of the size of the patient Any opening greater than 4 mm in diameter may cause cardiac failure, one of lesser diameter may thrombose Nevertheless, slight variation in the size of the anastomosis is almost inevitable Consequently it requires great experience to obtain uniform results Obviously the operation must be performed on the side of the descending aorta Therefore the operation is difficult in a patient with a right aortic arch *It appears that an anastomosis between the aorta and the pulmonary artery* grows better than a Blalock-Taussig anastomosis Indeed, in a number of instances the diameter of the opening has increased in size This is probably because greater stress and strain are placed on the aorta, as it grows, than on the pulmonary artery Hence there is real danger that the anastomosis may become too large and transmit the high pressure in the aorta to the pulmonary artery and consequently injure the pulmonary vascular bed

The insertion of a graft as formerly advocated by Gross in this country and commonly done in the U S S R , eliminates the sacrifice of the subclavian artery This technique, however, doubles the length of the operation and increases the risk of operation Although notching of the ribs on the side on which the subclavian artery is sacrificed is relatively common, this does no harm To the best of the author's knowledge, only one patient in 2,000 has experienced any serious complications from the ligation of the subclavian artery, therefore the insertion of a graft is indicated only in the rare instances when no vessel is obtainable

Direct attack and the excision of the stenotic area, as advocated by Brock, is possible only if a tetralogy of Fallot is associated with a valvular pulmonary stenosis or if there is a fair-sized infundibular chamber If there is an infundibular stenosis, a sufficient amount of muscle must be removed so as to prevent subsequent constriction from scar formation and, at the same time, the opening must

be sufficiently small to protect the lungs from the high pressure in the right ventricle which is secondary to the over riding of the aorta In the rare instance in which a tetralogy of Fallot has only a valvular pulmonary stenosis, care must be taken not to convert the malformation into an Eisenmenger complex Such an operation may be fatal, as the patient lacks the protective pulmonary hypertension characteristic of an Eisenmenger complex Indeed, unless the septal defect is closed excision of the stenotic area is not advisable

Corrective surgery is now possible Lillehei[6] was the first to demonstrate that with the aid of a pump and oxygenator it was possible to excise the stenotic area and close the ventricular defect and thereby to restore the heart and circulation to normal Such an operation is probably the ideal for adults and for children who have nearly attained their growth

For infants and young children with severe pulmonary stenosis or pulmonary atresia, in whom the pulmonary artery is but one third to one half as large as the aorta, it seems doubtful that the right ventricle, the pulmonary orifice, and the pulmonary artery will expand to normal size and grow normally with the child When a patch is necessary, it must be made of the proper size for the patient It is manifestly impossible to insert the patch of suitable size for an adult into a tiny infant, hence there is real danger that total correction in infancy or early childhood may leave the individual with an underdeveloped right ventricle and pulmonary stenosis by the time he reaches adolescence

Furthermore, recent experience has shown that a previous Blalock anastomosis does not greatly increase the risk of operation for total correction Moreover, Ferencz[7] studies on the pulmonary vascular bed have shown that, whereas most infants with anoxemia and decreased pulmonary blood flow have multiple thrombi in their lungs, patients with an adequate anastomosis who have died of unrelated causes have a normal pulmonary vascular bed Therefore the child who has had a previous anastomosis may be better able to adjust to a corrective procedure than the patient who has *not* had a previous operation

The indications for operation vary with the age of the patient and the severity of the pulmonary stenosis A baby with a tetralogy of Fallot who has a small heart but no murmur, and excessively clear lung fields, may be in real danger of dying from anoxemia, even though he has no polycythemia and but slight cyanosis Such infants frequently suffer from repeated attacks of paroxysmal dyspnea If the infant ceases to gain weight and the [illegible] mal dyspnea [illegible] operation is [illegible] the infant is gaining weight and doing well, it

is wise to postpone operation, as the risk of operation is less and the results are better in childhood than in infancy

The less severe the pulmonary stenosis, the louder is the systolic murmur Therefore, if the infant has a loud systolic murmur, early operation is seldom necessary *Indeed, if the attacks of paroxysmal dyspnea can be promptly relieved* by placing the infant on his abdomen in the knee chest position, it is usually safe to postpone operation Such attacks are most severe as the ductus arteriosus un dergoes obliteration and prior to the development of polycythemia

In brief, the decision concerning operation must be influenced by the fre quency and severity of these attacks and by whether the patient is losing ground or improving The determination of the arterial oxygen saturation may be of aid If it is only 30 per cent when the infant is crying, there is no cause for con cern If it is under 20 per cent, the condition is serious, and under 10 per cent is dangerously low

The persistence of attacks of paroxysmal dyspnea or loss of consciousness in *childhood is an indication for operation If the child s exercise tolerance is ex* tremely limited, that too is an indication for operation The early development of marked polycythemia is also an indication for operation, as when the hemato-crit reading reaches 70 per cent there is a real danger of thrombosis

Most parents are anxious for operation before the child starts school If, how ever, cyanosis is minimal and the red blood cell count is only 6 or 7 million cells per cu mm , and the limitation of exercise is slight, it may be wise to postpone surgery with the hope that only one operation will be necessary

If, however, the child is seriously incapacitated, it is wiser to operate while *the child is young and thus give him a normal childhood than to try to postpone* operation until total correction is indicated It is always advisable to operate *while the patient still has good tissue turgor and before the secondary changes* of the lungs due to long standing polycythemia become a serious problem The hemorrhagic diathesis is far more serious in adults than in children Even though a child may have a great reduction in platelets and a low blood fibrino gen, he seldom suffers from severe hemorrhage after the operation, whereas in the young adult hemorrhage in the early postoperative period may be a serious, even fatal, complication Therefore in 1960 it seems to the author advisable to perform a Blalock Taussig operation on an *infant or young child and to under* take total correction on older children with mild pulmonary stenosis or young *adults who have attained their growth*

The occurrence of a gross defect in the auricular septum may increase the strain on the circulation after an anastomotic procedure A number of patients

have developed considerable cardiac enlargement and a few have suffered from cardiac failure Nevertheless, a number of patients in whom cardiac catheterization indicated the presence of an auricular septal defect have had a smooth postoperative course and subsequently have experienced no difficulty from the additional defect Inasmuch as at the Johns Hopkins Hospital cardiac catheterization is not a routine procedure prior to operation, there are no figures available as to the incidence of auricular septal defects in our series of 1,500 patients with a tetralogy of Fallot

The occurrence of an auricular defect does, however, lessen the chance of an excellent result Therefore, when such an anomaly is known to exist, if possible it may be wise to postpone operation until such time as total correction may be of permanent benefit

PROGNOSIS

The prognosis varies with the severity of the pulmonary stenosis When the pulmonary stenosis is extreme or there is pulmonary atresia, the prognosis without operation is extremely poor In most instances closure of the ductus arteriosus renders the condition incompatible with life Therefore early operation is indicated

The tetralogy of Fallot has long been considered as the commonest malformation of the cyanotic group to be compatible with relative longevity Formerly all those who suffered from this malformation were able to receive great encouragement from a case reported by White and Sprague[1] of a man with a tetralogy of Fallot who lived for sixty years and was a concert violinist This man must have had a relatively mild pulmonary stenosis Few persons are as fortunate as he Today, however, persons with a pulmonary stenosis of such severity that it endangers or handicaps their lives can be greatly helped by surgery A Blalock Taussig operation gives a child a 75 per cent chance to enjoy a virtually normal life, at least, until adolescence Indeed, several women who have had this operation have borne normal children Once the patient has attained his growth, there is every reason to believe that total correction of the malformation not only will restore the circulation to normal but will be of permanent benefit Thus the prognosis is greatly improved

SUMMARY

The first known case of a tetralogy of Fallot was reported in 1671, but it was not until 1888 that Fallot clarified the syndrome which now bears his name

The four features which constitute the tetralogy of Fallot are pulmonary ste

nosis, dextroposition of the aorta, a high ventricular septal defect, and right ventricular hypertrophy

The pulmonary stenosis renders it difficult for the blood from the right ventricle to be pumped out to the lungs The dextroposition of the aorta means that some of the blood from the right ventricle which would normally go to the lungs for oxygenation is pumped into the aorta This leads to a reduction in the pulmonary blood flow and an increase in the systemic blood flow The pressure in the right ventricle is approximately the same as that in the systemic circulation The pressure in the pulmonary artery is low The work of the right ventricle is increased, there is right ventricular hypertrophy

The outstanding clinical features are cyanosis, dyspnea, a heart of normal size with a systolic murmur and a weak pulmonic second sound Cyanosis usually appears between three and six months of age The intensity of the cyanosis varies not only with the amount of reduced hemoglobin in the circulating blood but also with the amount of available hemoglobin

Cyanosis may be absent at birth The infant gains slowly and frequently suffers from attacks of paroxysmal dyspnea Cessation of weight gain and attacks of dyspnea progressing to loss of consciousness are ominous signs The infant is in danger of dying from anoxemia even though the heart is normal in size and there is no murmur

Cyanosis usually appears during the first year of life Compensatory polycythemia develops as the collateral circulation becomes established

Polycythemia gradually increases and may become extreme With long standing polycythemia the blood platelets and the blood fibrinogen become reduced

There is usually cyanosis and clubbing of the extremities The child frequently squats when tired

The heart is normal in size The intensity of the systolic murmur is inversely proportional to the severity of the pulmonary stenosis The pulmonic second sound is reduced in intensity but is usually audible

The x ray shows a small heart with a concave curve at its base to the left of the sternum and decreased hilar markings A right aortic arch occurs in 25 per cent of these patients

The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy

The circulation time is short

The oxygen saturation of the arterial blood is low and falls still further with exercise

Cardiac catheterization shows that the pressure in the right ventricle approaches systemic pressure and that the pressure in the pulmonary artery is low There is evidence of both a left to-right and a right to-left shunt

Angiocardiography may aid in the demonstration of the dextroposition of the aorta

Selective angiocardiography is of great value in the visualization of the outflow tract of the right ventricle

Diagnosis is based on the size and the contour of the heart and on the electrocardiographic findings of a right axis deviation and right ventricular hypertrophy in a cyanotic child who gives a history of paroxysmal dyspnea and who squats when tired

The condition requires differentiation from other malformations with pulmonary stenosis and a venous arterial shunt namely, pulmonary stenosis with an intact ventricular septum and a patent foramen ovale, a single ventricle, tricuspid atresia, a truncus arteriosus, complete transposition of the great vessels, occasionally from primary pulmonary hypertension or an Eisenmenger complex, and rarely from Ebstein s anomaly of the tricuspid valve

The most common associated anomaly is a gross defect in the auricular septum When this occurs the condition is known as a pentalogy of Fallot Occasionally there is but a single pulmonary artery and even more rarely the pulmonary valves are so abnormal that there is pulmonary insufficiency

The common complications are cerebral thrombosis, brain abscess, and subacute bacterial endocarditis Hemoptyses and hematemeses are rare complications, but do occur

Medical treatment is directed to the relief of paroxysmal dyspnea, to the prevention of infection and to the prevention of thrombosis For the last reason, prior to operation high fluid intake is important

Surgical treatment is of great benefit Any operation which increases the circulation to the lungs without raising the pressure with which the blood is ejected into the lungs is of great help A Blalock Taussig operation gives excellent results in childhood The ideal age for operation is from eight to twelve years, 90 per cent of the children in this age group who obtained good results from operation have maintained their benefi for the ensuing ten years

Closure of the ventricular defect and relief of the pulmonary stenosis by means of open heart surgery restores the circulation to normal The operation is performed with remarkable success in older individuals Not only is it more difficult in young children, but also there is real danger that, if the pulmonary orifice and pulmonary artery are abnormally small, the right side of the heart

nosis, dextroposition of the aorta, a high ventricular septal defect, and right ventricular hypertrophy

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will not grow in proportion to the growth of the child Were this to happen, the patient would be left with a pulmonary stenosis and an intact ventricular septum Therefore it seems wise to the author to reserve total correction until the patient has nearly attained his growth

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Courtesy of Dr J ... and ... the ...

FIGURE VII–1 Origin of both great vessels from the right ventricle and both of normal size

also arises from the right ventricle The pulmonary orifice is usually *stenosed* and the pulmonary valve may be bicuspid The left ventricle *lacks its normal* outflow tract the only way for blood to leave the left *ventricle is through* a ventricular septal defect Hence a ventricular septal defect is an *integral part* of the malformation Indeed, the size of the ventricular defect determines the ease with which blood can be pumped *out of the left* ventricle *and hence* the amount of work required of that chamber

The origin of the aorta from the *right ventricle makes* it the systemic ventricle, hence there is right ventricular hypertrophy *but the* pulmonary stenosis protects the lungs Unless the ventricular *defect is large*, there is difficulty in the expulsion of blood from the left ventricle, *hence it, too*, undergoes dilatation and hypertrophy The foramen ovale is closed and the ductus arteriosus undergoes normal obliteration Figure VII–2 illustrates this type of malformation

COURSE OF THE CIRCULATION

During fetal life the course of the circulation is grossly altered by the abnormal position of the aorta Most of the blood from the right auricle which

CHAPTER VII

ORIGIN OF BOTH GREAT VESSELS FROM THE RIGHT VENTRICLE

THIS malformation represents a further rotation of the aorta than that which occurs in the tetralogy of Fallot The aorta is so far dextroposed that it arises entirely from the right ventricle The pulmonary artery is usually stenotic and in most instances it occupies its normal position, as it does in the tetralogy of Fallot, thus it, too, arises from the right ventricle Between 1948 and 1959 we have studied five cases in which both great vessels arose from the right ventricle [1] Occasionally the stenosed pulmonary artery lies posterior and *slightly to the right of the aorta but arises entirely from the right ventricle* Under such circumstances the relation of the two great vessels is that of a corrected transposition, nevertheless, both great vessels arise from the right ventricle This malformation was known to Maude Abbott [2] Lev and Saphir,[3] in a report of six cases of transposition of the arterial trunk, described three such cases Witham[4] in 1957 reported the clinical and pathological findings in four cases and reviewed the literature

*If the pulmonary artery is atretic, it makes no difference whether it lies an*terior or posterior to the aorta When an atretic pulmonary artery arises from the left ventricle, the condition constitutes a transposition of the great vessels, therefore the condition is discussed in Chapter x, Section c

This malformation may also occur with a pulmonary artery of normal size (see Figure vii-1) *Under such circumstances there is adequate circulation to the* lungs, but pulmonary congestion usually occurs early, as a large volume of blood is ejected to the lungs under systemic pressure and great difficulty is encountered in the expulsion of blood from the left ventricle When the ventricular septal defect is relatively large, the condition may be compatible with life for a number of years Under such circumstances the malformation *simulates the Eisenmenger* complex but differs from it in that cyanosis dates from birth and the heart undergoes slow progressive enlargement

The present discussion is limited to the malformation as it occurs with pulmonary stenosis

NATURE OF THE MALFORMATION

The aorta arises entirely from the right ventricle and the pulmonary artery



Courtesy of Dr J E Edwards and the Mayo Clinic

FIGURE VII-1 Origin of both great vessels from the right ventricle and both of normal size

also arises from the right ventricle The pulmonary orifice is usually stenosed and the pulmonary valve may be bicuspid The left ventricle lacks its normal outflow tract the only way for blood to leave the left ventricle is through a ventricular septal defect Hence a ventricular septal defect is an integral part of the malformation Indeed, the size of the ventricular defect determines the ease with which blood can be pumped out of the left ventricle and hence the amount of work required of that chamber

The origin of the aorta from the right ventricle makes it the systemic ventricle, hence there is right ventricular hypertrophy but the pulmonary stenosis protects the lungs Unless the ventricular defect is large, there is difficulty in the expulsion of blood from the left ventricle, hence it, too, undergoes dilatation and hypertrophy The foramen ovale is closed and the ductus arteriosus undergoes normal obliteration Figure VII-2 illustrates this type of malformation

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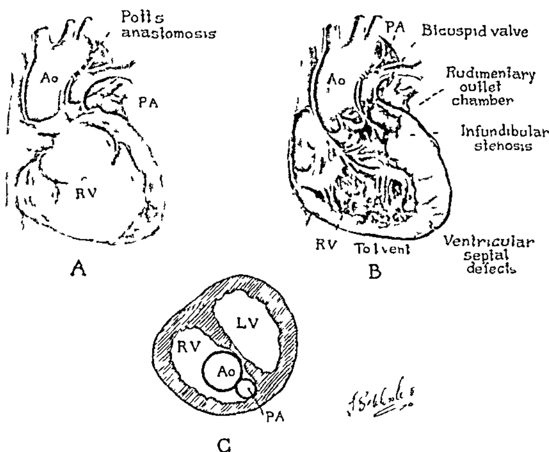


FIGURE VII–2 Origin of both great vessels from the right ventricle and an abnormally small pulmonary artery

flows into the right ventricle is pumped out through the aorta to the body and is returned by the superior vena cava and the inferior vena cava to the right auricle. Some of the blood from the right ventricle is pumped out through the pulmonary artery to the lungs, and some blood may flow through the ductus arteriosus to the lungs. All the blood which goes to the lungs is returned in the normal manner to the left auricle. In addition to the blood returned from the lungs to the left auricle, some blood from the right auricle flows through the foramen ovale to the left auricle. All the blood in the left auricle flows into the left ventricle. Here difficulty is encountered in the ejection of blood from the left ventricle, because the only way for blood to be pumped out of the left ventricle is through the septal defect to the right ventricle. This increases the work of the left ventricle. Hence at birth the two ventricles are of approximately equal thickness (see Figure VII–3).

After birth the course of the circulation remains essentially the same The blood from the right auricle flows into the right ventricle and is pumped out into the aorta and also into the pulmonary artery The expansion of the lungs lowers the pulmonary pressure and increases the volume of venous blood which flows through the pulmonary artery and also that which flows from the aorta through the ductus arteriosus to the lungs All the blood which flows to the lungs is oxygenated and returned by the pulmonary veins to the left auricle The increased circulation to the lungs increases the volume of blood returned to the left auricle and tends to close the foramen ovale The blood in the left auricle flows into the left ventricle The increased volume of blood which is returned to the left ventricle increases the volume of blood which must pump through the septal defect to the right ventricle and thence out into the aorta and the pulmonary artery The blood which flows to the body is returned in the normal manner by the superior vena cava and the inferior vena cava to the right auricle So the cycle continues The course of the circulation is shown in Diagram VII–1

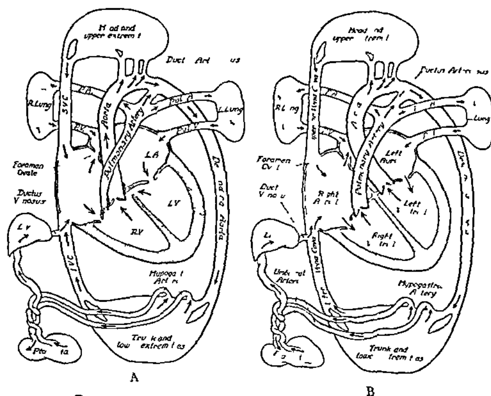


FIGURE VII–3 Fetal circulation (A) Origin of both great vessels from the right ventricle and (B) normal h art

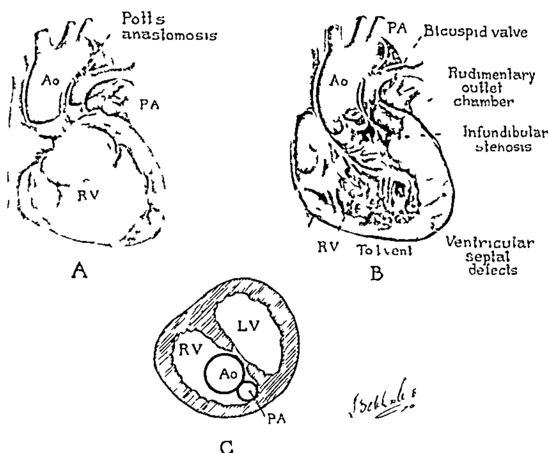


FIGURE VII 2 Origin of both great vessels from the right ventricle and an abnormally small pulmonary artery

flows into the right ventricle is pumped out through the aorta to the body and is returned by the superior vena cava and the inferior vena cava to the right auricle Some of the blood from the right ventricle is pumped out through the pulmonary artery to the lungs, and some blood may flow through the ductus arteriosus to the lungs All the blood which goes to the lungs is returned in the normal manner to the left auricle In addition to the blood returned from the lungs to the left auricle, some blood from the right auricle flows through the foramen ovale to the left auricle All the blood in the left auricle flows into the left ventricle Here difficulty is encountered in the ejection of blood from the left ventricle, because the only way for blood to be pumped out of the left ventricle is through the septal defect to the right ventricle This increases the work of the left ventricle Hence at birth the two ventricles are of approximately equal thickness (see Figure VII-3)

Diagram VII-1

Origin of both great vessels from the right ventricle and a small pulmonary artery

The essential feature of this malformation is that the aorta is transposed and arises from the right ventricle the pulmonary artery is stenosed and it too, arises from the right ventricle In order for the blood to escape from the left ventricle there must be a ventricular septal defect.

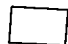
The blood from the right auricle flows into the right ventricle Most of the blood in the right ventricle is pumped out through the aorta to the systemic circulation and is returned by the superior vena cava and the inferior vena cava to the right auricle Inasmuch as there is pulmonary stenosis, only a small amount of blood is pumped through the pulmonary artery to the lungs where it is oxygenated and returned to the left auricle So long as the ductus arteriosus remains open, some blood flows through the ductus arteriosus to the lungs This increases the circulation to the lungs and also increases the volume of blood returned to the left auricle and the left ventricle

Since both the aorta and the pulmonary artery arise from the right ventricle the only way for the blood to leave the left ventricle is through the septal defect to the right ventricle If this is small the left ventricle has difficulty in emptying itself hence there is also left ventricular hypertrophy

Clinical diagnosis Inasmuch as the aorta arises from the right ventricle the infant shows persistent cyanosis he does not suffer from attacks of paroxysmal dyspnea but may have episodes of loss of consciousness The heart is enlarged There is absence of fullness of the pulmonary conus Viewed in the left anterior-oblique position the right ventricle is enlarged there may or may not be enlargement of the left ventricle The electrocardiogram shows a right axis deviation and evidence of right ventricular strain" Death results from anoxemia or cardiac failure.

Diagram VII–1

Head and upper extremities
R Lung
PA
S.V.C.
PV
Aorta
Pul A.
L Lung
Pul V
L.A.
Pul Art.
R.A.
L.V.
R.V.
I.V.C.
Descending Aorta
Trunk and lower extremities


Arterial blood (fully saturated)


Small admixture of venous blood
No visible cyanosis


Venous and arterial blood
Cyanosis visible


Venous blood

X RAY AND FLUOROSCOPIC FINDINGS

The heart is enlarged, especially to the left There is no fullness in the region of the pulmonary conus The cardiac shadow extends downward and outward from a narrow base (see Figure VII-4) The lungs are clear and the hilar markings are inconspicuous

In the left anterior-oblique position both ventricles are seen to be enlarged The right ventricle extends out toward the chest wall and the shadow of the left ventricle extends posteriorly far beyond the spinal column, as seen in Figure VII-5 The small size of the pulmonary artery renders the pulmonary window abnormally clear In addition, the origin of the aorta from the right ventricle causes the pulmonary window to be unusually wide Consequently the pulmonary window is both wide and clear

Examination in the right anterior-oblique position contributes little

ELECTROCARDIOGRAPHIC FINDINGS

The standard leads show a right axis deviation and the unipolar precordial leads show evidence of right ventricular hypertrophy and the pattern of so-



FIGURE VII-4 Origin of both great vessels from the right ventricle and pulmonary stenosis (same patient as in Figures VII-5 6, 7) Infant

PHYSIOLOGY OF THE MALFORMATION

In this malformation the right ventricle pumps the blood into the aorta, therefore the systolic pressure in the right ventricle is the same as that in the systemic circulation The occurrence of pulmonary stenosis, however, protects the lungs and the pulmonary pressure is low The combined blood flow through the stenosed pulmonary artery and the ductus arteriosus is usually sufficient for life Nevertheless, only a small volume of blood is directed to the lungs and a large volume of mixed venous and arterial blood flows to the body, where it gives up more of its oxygen Hence a small volume of oxygenated blood is mixed with a large volume of venous blood which is extremely low in oxygen content It follows that the oxygen content of the mixed venous and arterial blood is abnormally low

CLINICAL FINDINGS

Cyanosis appears early, it usually dates from birth The infant frequently suffers from "spells" during which the cyanosis increases in intensity The spell may be long and severe and progress to loss of consciousness

Polycythemia develops early and may become extreme The red blood cell count may reach 9 or 10 million cells per cu mm, the level of the available hemoglobin and the hematocrit reading are proportionally high

Dyspnea is usually severe but *attacks of paroxysmal dyspnea* seldom occur

Barrel shaped chest deformity is the rule

The lungs remain clear, as the pulmonary blood flow is meager

Growth and development are severely retarded

Exercise tolerance is markedly restricted Children with this malformation seldom learn to walk before three years of age, thereafter they squat when tired

The liver may be enlarged and pulsations may be detectable at its margin

Edema and ascites are late manifestations of cardiac failure

CARDIAC FINDINGS

The heart is enlarged *A systolic murmur* and a *thrill* are usually present The *second sound* at the base of the heart to the left of the sternum is accentuated owing to the transposed aorta A *gallop rhythm* may be present

Cardiac failure occurs early It is primarily right sided failure with enlargement of the liver and edema of the extremities

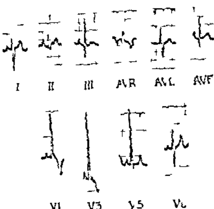


FIGURE VII-6 Origin of both great vessels from the right ventricle and pulmonary stenosis (same patient as in Figure VII-4) Infant

the catheter usually enters the aorta with ease If the pulmonary artery can be catheterized, the pulmonary pressure is low The oxygen content of the blood in the aorta and in the pulmonary artery is virtually the same

Angiocardiography shows that the aorta fills simultaneously with a small pulmonary artery It is of interest that the case which illustrates this malformation was misdiagnosed because it was thought that the aorta was not transposed The aorta did *not* appear to lie as far anteriorly as is usual when the aorta is transposed Nevertheless, careful examination of the aorta relative to the left ventricle in the lateral films taken after the left ventricle has filled suggests that the aorta lies anterior to the left ventricle (see Figure VII-7) Selective angiocardiography, with dye placed in the pulmonary artery, should clearly demonstrate the left auricle, the left ventricle, and the anterior position of the aorta

DIAGNOSIS

The diagnosis is based upon the finding of persistent cyanosis, which appears at an early age and is associated with severe dyspnea, absence of attacks of paroxysmal dyspnea the occurrence of episodes of deepening cyanosis which often [illegible]

The ab[illegible]

cardiac

[illegible] the heart to the left of the sternum show that the pulmonary artery is diminutive, misplaced, or absent In the left anterior-oblique position both ventricles are seen to be greatly enlarged and the pulmonary window is wide and clear There is electrocardiographic evidence of extreme right ventricular hypertrophy



Left anterior oblique position



Right anterior oblique position

FIGURE VII–5 Origin of both great vessels from the right ventricle and pulmonary stenosis (same patient as in Figure VII–4) Infant

called right ventricular 'strain The P waves are often high and peaked (see Figure VII–6)

SPECIAL TESTS

The circulation time (arm to tongue) is short

The oxygen saturation of the arterial blood is low and falls still further with exercise

Cardiac catheterization reveals systemic pressure in the right ventricle and

DIFFERENTIAL DIAGNOSIS

This malformation requires differentiation from a defective development of the right ventricle with an intact ventricular septum and pulmonary stenosis, from a tetralogy of Fallot with severe pulmonary stenosis or functional pulmonary atresia, from truncus arteriosus with reduced pulmonary blood flow, and from a complete transposition of the great vessels with pulmonary stenosis

Defective development of the right ventricle with an intact ventricular septum is readily confused with the malformation under discussion When there is a defective development of the right ventricle but a normally placed aorta, the infant does not suffer from attacks of paroxysmal dyspnea If, however, an angiocardiography is performed, although the pulmonary artery will be seen to arise from the right ventricle, the aorta will not be visualized until after the left auricle and the left ventricle have filled

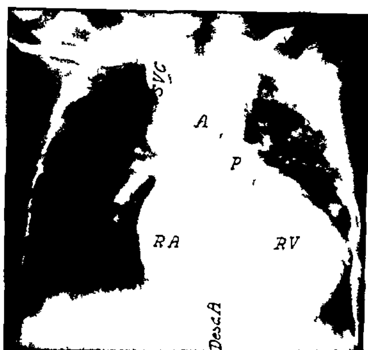
A tetralogy of Fallot with anatomical or functional pulmonary atresia causes the heart to remain small and in the left anterior-oblique position it appears to be globular In contrast to this, when both great vessels arise from the right ventricle, that chamber is always enlarged, if the duration of life permits, the left ventricle will also become enlarged because of the difficulty in the expulsion of blood from the left ventricle

Truncus arteriosus in which both ventricles are normally formed and the circulation to the lungs is through the enlarged bronchial arteries, is differentiated by x ray examination The *truncus* is usually larger than the aorta and the aortic knob lies at an abnormally high level The heart is also relatively larger and extends out as a shelf to the left of the sternum The infant usually does better than in the malformation under discussion

Complete transposition of the great vessels combined with pulmonary stenosis may in early infancy closely resemble the malformation under discussion When the great vessels are transposed, the vascular markings extend to the periphery of the lung fields

TREATMENT

Treatment is unsatisfactory A systemic pulmonary anastomosis increases the volume of venous blood directed to the lungs but there is usually such great difficulty in the expulsion of blood from the left ventricle that the infant dies from pulmonary edema The creation of an auricular defect may lessen this difficulty but after operation the strain on the circulation is so great that the child dies from cardiac failure



Simultaneous visualization of the aorta and the pulmonary artery



Superimposition of the aorta and *pulmonary artery*

FIGURE VII–7 Origin of both great vessels from the right ventricle and pulmonary stenosis (same patient as in Figure VII–4) Infant

also from a complete transposition of the great vessels with pulmonary stenosis

Treatment is unsatisfactory because any increase in the circulation to the lungs increases the volume of blood returned to the left ventricle Usually the difficulty in the expulsion of the blood from the left ventricle is so great that the infant dies of pulmonary edema Without operation the child usually dies of anoxemia or cardiac failure Therefore, either with or without operation the prognosis is poor Most patients die in early childhood

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3 Lev M and O Saphir Transposition of the large vessels J Tech Methods 17 1 6-16 1937

4 Witham C A Double outlet right ventricle Am Heart J 53 928-939 1957

Relief of the pulmonary stenosis under direct vision may improve the circulation to the lungs, but, if the pulmonary artery is widely opened, the blood will be pumped to the lungs under systemic pressure Furthermore, because of the difficulty in the expulsion of blood from the left ventricle, the increased circulation to the lungs usually precipitates pulmonary edema

PROGNOSIS

Prognosis is poor Most patients die in infancy or early childhood Nevertheless, the author knows of three patients who lived to adult life and died at operation A systemic pulmonary anastomosis combined with the creation of an auricular defect may lessen the incapacity but frequently leads to acute pulmonary edema or intractable cardiac failure

SUMMARY

The origin of both great vessels from the right ventricle, with or without pulmonary stenosis, is a very serious malformation The fact that the aorta arises from the right ventricle means that venous blood is pumped directly into the systemic circulation In the presence of pulmonary stenosis little blood reaches the lungs for oxygenation In the absence of pulmonary stenosis blood is ejected to the lungs under systemic pressure, furthermore, owing to the difficulty in the expulsion of blood from the left ventricle, the pressure in the left auricle is increased This also leads to pulmonary hypertension Consequently pulmonary hypertension is severe The condition soon leads to pulmonary edema

The outstanding clinical findings are intense cyanosis, dyspnea, and absence of attacks of paroxysmal dyspnea The child squats when tired

The cardiac findings are similar to those of a tetralogy of Fallot with great over riding of the aorta The pulmonic second sound is accentuated but not reduplicated The x ray shows moderate cardiac enlargement and a concave curve at the base of the heart The lung fields are usually clear Viewed in the left anterior oblique position both ventricles are enlarged and the pulmonary window is wide and clear

The electrocardiogram shows a right axis deviation and evidence of extreme right ventricular hypertrophy

Angiocardiography shows that both vessels are filled from the right ventricle

The condition requires differentiation from defective development of the right ventricle with an intact ventricular septum and pulmonary stenosis a tetralogy of Fallot, a truncus arteriosus with reduced pulmonary blood flow, and

also from a complete transposition of the great vessels with pulmonary stenosis

Treatment is unsatisfactory because any increase in the circulation to the lungs increases the volume of blood returned to the left ventricle Usually the difficulty in the expulsion of the blood from the left ventricle is so great that the infant dies of pulmonary edema Without operation the child usually dies of anoxemia or cardiac failure Therefore, either with or without operation the prognosis is poor Most patients die in early childhood

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CHAPTER VIII

TRICUSPID ATRESIA AND DEFECTIVE DEVELOPMENT OF THE RIGHT VENTRICLE

TRICUSPID ATRESIA *means that there is no tricuspid orifice, hence the* blood cannot reach the right ventricle in the normal manner, consequently *the right ventricle is always abnormally small.* For this reason the two conditions are usually part and parcel of the same malformation. The right ventricle may be absent or it may persist as a blind sac which does not communicate with the circulation. Sometimes the primitive outflow tract develops as a small chamber which receives its blood from the left ventricle through a defect in the ventricular septum.[1] The great vessels may or may not be transposed. In all instances it is the diminutive size of the right ventricle which offers the clue to the nature of the underlying malformation. The small size of the right ventricle can be seen by x-ray and fluoroscopy and is confirmed by the electrocardiographic finding of a left ventricular preponderance in the precordial leads.

Section A is concerned with tricuspid atresia combined with defective development of the right ventricle in which the great vessels are normally placed. Section B is concerned with tricuspid atresia when there is a small right ventricle from which the aorta arises.

A. *Tricuspid Atresia Combined with Defective Development of the Right Ventricle and with Normally Placed Great Vessels*

NATURE OF THE MALFORMATION

The essential feature of this malformation is the atresia of the tricuspid valve, its orifice is represented by a dimple in the base of the right auricle. In addition, there is defective development of the right ventricle. *The right ventricle may* be absent, there may be a rudimentary chamber which has no demonstrable connection with the circulation, or in rare instances there may be extreme *hypoplasia* of the tricuspid valve and the right ventricle may exist as a diminutive chamber which is filled with a blood clot. In each of these conditions the right ventricle fails to function as an integral part of the heart. When the tricuspid valve is atretic or markedly hypoplastic and the right ventricle is a non-functioning chamber, there is inevitably some malformation of the pulmonary orifice. A

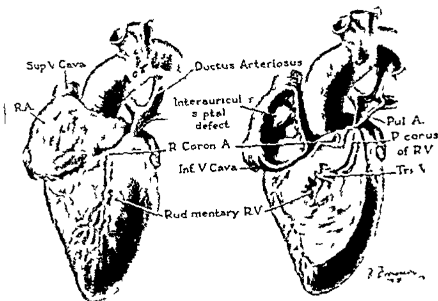


FIGURE VIII-1 Tricuspid atresia a non functioning right ventricle and a gross auricular septal defect (same patient as in Figure VIII-8) Infant

The diminutive size of the right ventricle caused marked distortion of the specimen on fixation

functioning pulmonary artery cannot be given off a non functioning chamber It follows that the pulmonary artery is either atretic or occupies an abnormal position Usually there is pulmonary atresia and the blood reaches the lungs through the ductus arteriosus, as shown in Figure VIII-1

In some instances the primitive bulbus cordis develops into an outflow chamber which receives its blood from the left ventricle through a defect in the ventricular septum Under such circumstances the abnormality concerns the tricuspid valve and the development of the right ventricle, the pulmonary artery is usually normally formed and of relatively normal size Furthermore, the circulation to the lungs is proportionately better when there is a small right ventricle than when the right ventricle is non functioning Such is the usual structure of the heart in patients with tricuspid atresia who survive for more than two years, the condition may be compatible with life for a number of years Figure VIII-2 illustrates this type of anomaly in a four year-old child, and Figure VIII-3 is a drawing of the heart of a patient who died at operation at thirty five years of age

The atresia of the tricuspid orifice and the non functioning right ventricle

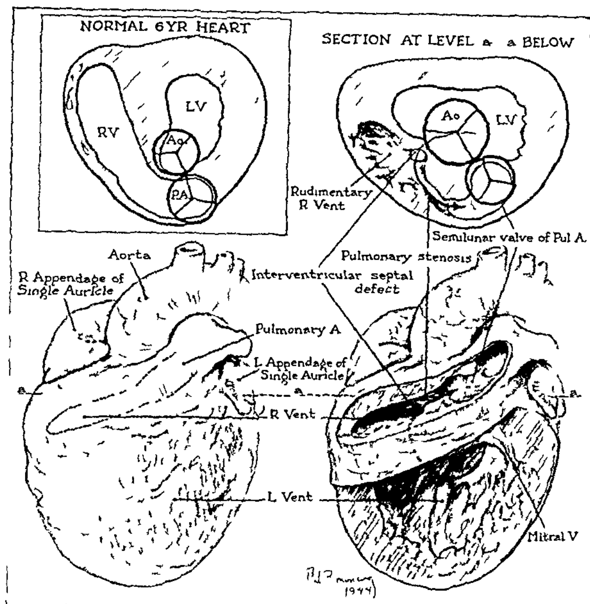


FIGURE VIII-2 Tricuspid atresia and a rudimentary right ventricle which receives blood from the left ventricle through a small ventricular septal defect *(same patient as in Figure VIII-11) Child*

mean that the blood which enters the right auricle through the superior and inferior venae cavae cannot leave that chamber in the normal manner The blood from the right auricle must escape by way of the left auricle This renders inevitable some defect in the auricular septum The foramen ovale may be normal in structure and covered by a valve which is not completely sealed, or there may be a gross defect in the auricular septum In either case there are two auricles and one ventricle, which constitutes a triloculate heart In some cases the auricular septum is entirely lacking except for a few strands of tissue Under

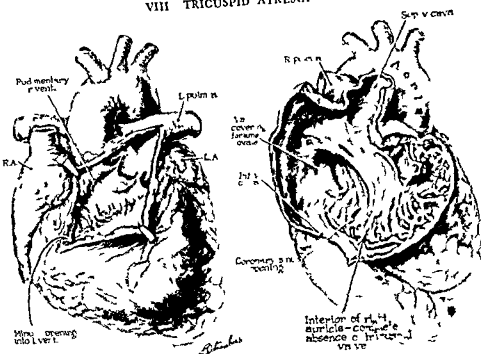


FIGURE VIII-3 Tricuspid atresia with the pulmonary artery arising from a rudimentary right ventricle Adult

such circumstances the two auricles function as a single chamber There is but one auricle and one ventricle, functionally there is a biloculate heart In order to make a clinical diagnosis it is necessary first to establish the diagnosis of tricuspid atresia and a diminutive or absent right ventricle and then to analyze the condition within the auricles

COURSE OF THE CIRCULATION

During fetal life the foramen ovale and the ductus arteriosus are normally patent Inasmuch as the tricuspid orifice is atretic and the right ventricle does not function all the blood from the right auricle must flow through the foramen ovale into the left auricle and thence to the left ventricle The left ventricle pumps the blood through the aorta to the body of the fetus and through the ductus arteriosus into the branches of the pulmonary artery and thence to the lungs Inasmuch as the lungs do not function during fetal life, the normal flow of blood to the lungs is minimal, hence the absence of the right ventricle places little strain upon the fetal circulation The heart at birth is of normal size Figure VIII-4 shows the course of the fetal circulation when there is tricuspid atresia and pulmonary atresia Figure VIII-5 shows the course of the fetal circulation

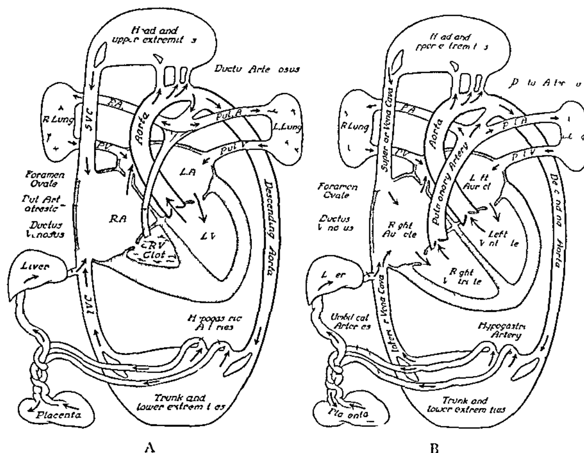


FIGURE VIII-4 Fetal circulation (A) Non functioning right ventricle and a well formed auricular septum and (B) normal heart

when in spite of the tricuspid atresia the pulmonary artery arises from the infundibular chamber of the right ventricle

After birth some circulation to the lungs must be established, usually this is by way of the ductus arteriosus It is, of course, possible to have other pathways, such as a transposed pulmonary artery or a truncus arteriosus Such conditions, however, represent additional anomalies (discussed in Section B and in Chapter XIV, respectively), whereas the ductus arteriosus represents the persistence of a normal fetal pathway

As long as the ductus arteriosus remains patent, the blood from the aorta passes through it to the pulmonary artery The pulmonary artery proximal to the ductus arteriosus usually persists as a small non functioning vessel The main branches of the pulmonary artery beyond the ductus arteriosus are normally developed The course of the circulation is shown in Diagram VIII-1

All the blood which enters the right auricle flows into the left auricle and thence to the left ventricle Therefore, the blood in the left ventricle is a mix

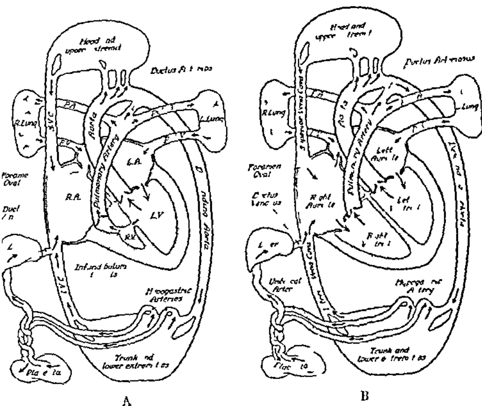


Figure viii–5 Fetal circulation. (A) Tricuspid atresia and stenosis of the infundibulum and (B) normal heart

ture of oxygenated and venous blood The blood in the left ventricle is pumped out through the aorta to the systemic circulation, blood from the aorta also flows through the ductus arteriosus to the pulmonary artery and thence to the lungs The oxygenated blood from the lungs is returned by the pulmonary veins to the left auricle The blood from the systemic circulation is returned by the superior vena cava and the inferior vena cava to the right auricle, thence it flows to the left auricle There the cycle starts again

Absence of the auricular septum does not greatly alter the course of the circulation The blood from the superior vena cava and the inferior vena cava enters the right side of the common auricle, and that from the pulmonary veins enters the left side All the blood from both auricles flows through the mitral valve to the left ventricle and out by way of the aorta to the systemic circulation and through the ductus arteriosus to the pulmonary arteries and the lungs, as shown in Diagram viii–2

When the right ventricle remains as a rudimentary chamber which receives

Diagram VIII–1

Head and upper extremities

Ductus Arteriosus

R Lung

SVC

PA

Aorta

Pul A

L Lung

PV

Pul V

A

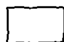
LV

Descending Aorta

Remnant of Pul Ar

IVC

Trunk and lower extremities



Arterial blood (fully saturated)



Venous and arterial blood
Cyanosis visible

Small admixture of venous blood
No visible cyanosis



Venous blood

Diagram VIII-1

Marked hypoplasia of the tricuspid orifice and a non functioning right ventricle combined with a well formed auricular septum pulmonary atresia and a patent ductus arteriosus

In this malformation there are two auricles and a single ventricle the left The right ventricle, even if present is diminutive in size and does not function The tricuspid valve is atretic or markedly hypoplastic In addition, there is pulmonary atresia hence the only way for the blood to reach the lungs is by way of the ductus arteriosus The auricular septum is well formed but there is either a small defect or a functionally patent foramen ovale

The abnormality of the tricuspid valve and the diminutive right ventricle prevent the normal flow of blood from the right auricle to the right ventricle Whatever blood does enter the right ventricle cannot escape and becomes clotted The only way for the blood to circulate is from the right auricle, through the foramen ovale, into the left auricle Thence it flows into the left ventricle and is pumped out by way of the aorta to the systemic circulation and through the ductus arteriosus to the lungs The blood from the systemic circulation is returned in the normal fashion by the superior vena cava and the inferior vena cava to the right auricle and thence to the left auricle Inasmuch as there is pulmonary atresia the pressure in the pulmonary circulation is low and blood will flow from the aorta through the ductus arteriosus to the lungs This is the only way by which the blood can reach the lungs The blood from the lungs is returned in the normal fashion by the pulmonary veins to the left auricle

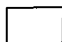
The volume of blood shunted through the ductus arteriosus is small Consequently only a small amount of blood reaches the lungs for oxygenation and a correspondingly small amount of oxygenated blood is returned to the left auricle Hence the pressure in the left auricle remains low This aids the flow of blood from the right auricle to the left auricle The small amount of oxygenated blood returned to the left auricle and the large volume of venous blood shunted from the right auricle to the left auricle produce cyanosis

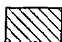
As the infant grows and the ductus arteriosus undergoes normal obliteration the supply of blood to the lung decreases Cyanosis becomes progressively more intense With the closure of the ductus arteriosus the condition usually becomes incompatible with life Death commonly occurs between the ages of three and five months

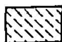
Clinical diagnosis is made by x ray or fluoroscopic examination The shape of the heart is characteristic The absence of the right ventricle causes the left ventricle to enlarge In the anterior posterior view there is absence of the shadow normally cast by the pulmonary conus In the left anterior-oblique position the enlargement of the heart is seen to be due to the left ventricle the right ventricle does not project forward beyond the margin of the aorta The pulmonary window is unusually clear Murmurs if present, are systolic in time and are of no diagnostic aid The electrocardiogram shows a left axis deviation and evidence of left ventricular dominance

Diagram VIII–2

Head and upper extremities
Ductus Arteriosus
R Lung
S.V.C.
P.A.
Aorta
Pul. A.
L Lung
PV
Pul V
L.A.
R.A.
L.V.
Descending Aorta
Remnant of Pul Art
R.V.
I.V.C.
Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

DIAGRAM VIII–2

Tricuspid atresia and a non functioning right ventricle combined with a gross auricular septal defect pulmonary atresia and a patent ductus arteriosus

In this malformation in addition to the non functioning right ventricle and the pulmonary atresia there is such a gross defect in the auricular septum that the two auricles function as a single chamber Each auricle occupies its normal position There is either tricuspid atresia or marked hypoplasia of the tricuspid orifice

The blood from the right auricle cannot leave by its normal pathway it can escape only by way of the mitral orifice into the left ventricle The blood from the left ventricle is pumped out through the aorta to the systemic circulation and is returned by the superior vena cava and the inferior vena cava to the right auricle Thence it again flows into the left auricle and the left ventricle Inasmuch as there is pulmonary atresia, the pressure in the pulmonary circulation is low So long as the ductus arteriosus is patent [illegible]

[illegible] the lungs is returned by the pulmonary veins in the normal fashion to the left auricle

Thus the left auricle receives oxygenated blood from the lungs and venous blood from the systemic circulation this mixture of arterial and venous blood flows into the left ventricle and is pumped into the aorta The greater portion of the blood in the aorta is pumped into the systemic circulation only a relatively small volume of blood reaches the lungs for oxygenation It follows that only a small volume of oxygenated blood is returned to the left auricle where it is mixed with a large volume of venous blood received from the right auricle Cyanosis is intense As the ductus arteriosus closes the condition becomes incompatible with life

Cl[illegible] the[illegible] The right ventricle is [illegible] the left ventricle which does the work of both ventricles is enlarged In the anterior posterior view there is an absence of the fullness of the pulmonary conus In the left anterior-oblique position there is an absence of enlargement of the right ventricle The gross defect in the auricular septum increases the pressure in the right auricle and causes dilatation of the superior vena cava A systolic murmur may or may not be present such murmurs are of no diagnostic aid During the neonatal period the electrocardiogram may show a balanced axis subsequently the electrocardiogram will show a left axis deviation but the unipolar precordial leads show evidence of left ventricular dominance which is present at birth and conspicuous in V_1 as well as in V_5 and V_6

its blood from the left ventricle, the circulation is altered only by the volume of blood which is pumped from the left ventricle into the rudimentary right ventricle and thence through the pulmonary artery to the lungs In such cases, although the pulmonary blood flow is reduced, life is not dependent upon the patency of the ductus arteriosus Consequently this condition may be compatible with life for a number of years The course of the circulation is shown in Diagram VIII-3

PHYSIOLOGY OF THE MALFORMATION

The altered structure of the heart causes difficulty in the expulsion of blood from the right auricle and in the direction of blood to the lungs Consequently there is always a reduction in the pulmonary blood flow except when the malformation occurs in combination with a transposition of the great vessels (see Section B) The reduction in the pulmonary blood flow, combined with the admixture of venous and arterial blood in the left ventricle, causes a marked reduction in the oxygen saturation of the arterial blood The pulmonary stenosis reduces the pressure in the pulmonary artery, hence the malformation does not injure the lungs When there is pulmonary atresia, life is dependent on the patency of the ductus arteriosus or on the development of collateral circulation

CLINICAL FINDINGS

Cyanosis is persistent and usually dates from birth In this malformation only a small amount of blood reaches the lungs for oxygenation and therefore only a small amount of oxygenated blood is returned to the left auricle, where it is mixed with a large volume of venous blood which has been returned from the systemic circulation to the right auricle and thence to the left auricle This mixture of venous and arterial blood passes from the left auricle to the left ventricle and is pumped out into the aorta Therefore cyanosis is intense and of uniform distribution

Clubbing of the fingers and toes develops at an early age

Difficulty in feeding is a common complaint Most infants with this malformation gain weight very slowly *Failure to gain* is an ominous sign

The occurrence of attacks of paroxysmal dyspnea is usually extremely serious, such attacks occur as the ductus arteriosus undergoes obliteration Moreover, life is usually dependent upon the patency of the ductus arteriosus In this malformation there is real danger that the first attack of paroxysmal dyspnea will be the last

If the infant is able to establish sufficient circulation to the lungs, he out grows the attacks of paroxysmal dyspnea between eighteen months and two years of age, thereafter he starts to improve

Squatting is quite as common a habit for these children as for those with a tetralogy of Fallot Indeed, their exercise tolerance varies in a similar manner to that of patients with a tetralogy of Fallot and their general behavior is also closely similar

The blood pressure is narrow and may be difficult to obtain The pulses in the arm and the leg are of equal strength

The liver may or may not be enlarged Pulsations at the margin are frequently palpable Even when the liver is normal in size, the heart beat may be readily counted by placing one s hand against the margin of the liver in the anterior axillary line When such pulsations are palpable, the opening between the two auricles is small

CARDIAC FINDINGS

The heart is usually slightly enlarged Generally the apex thrust is palpable beyond the mid clavicular line

The quality of the second sound is remarkably pure Since there is only *one great* vessel, there can be no reduplication of the second sound It is to be emphasized that it is the *purity* of this sound, not the location, which is of significance The pulmonic second sound may be louder than the aortic second sound This does not mean that the sound is produced by the closure of the pulmonary valves It means only that the closure of the aortic valve is better heard to the left of the sternum than to the right

Murmurs may or may not be present The only abnormal openings which could cause murmurs are the ductus arteriosus and a defect in the auricular septum The defect between the auricles may be so large that the two auricles function as a single chamber The ductus *arteriosus* is not merely a connection between the two circulations but is the main pathway to the lesser circulation Inasmuch as the occurrence of a murmur depends upon the size of the openings and the relative pressure in the two circulations, there may or may not be a murmur For this reason murmurs are of no diagnostic importance

The structure of the auricular septum is adduced from the size of the liver and the presence or absence of pulsations at the margin of the liver

A well formed auricular septum when combined with a non functioning right ventricle, causes the heart to function as a triloculate one, that is, two auri

DIAGRAM VIII–3

Head and upper extremities

R Lung

L Lung

S.V.C.

PA

PV

Aorta

Pulmonary Artery

Pul A

Pul V

L.A.

R.A.

L.V.

R.V.

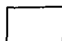
Descending Aorta

Infundibulum Stenosis

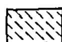
Defect in Interventricular Septum

I.V.C.

Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

Diagram VIII-3

Tricuspid atresia and a gross auricular septal defect combined with defective development of the right ventricle infundibular stenosis and a small ventricular septal defect

The essential features of this malformation are tricuspid atresia and defective development of the right ventricle Inasmuch as the tricuspid valve is atretic the blood must flow from the right auricle to the left auricle it follows that either the foramen ovale is held open by the high pressure in the right auricle or there is a defect in the auricular septum The only way for the blood to enter the right ventricle is from the left ventricle through a ventricular septal defect There may also be infundibular stenosis of the right ventricle which places an additional obstruction to the flow of blood through the right ventricle to the pulmonary artery

The blood flows from the right auricle through the defect in the auricular septum into the left auricle, where it meets the blood which is returned by the pulmonary veins to the left auricle This mixture of venous blood from the right auricle and oxygenated blood from the left auricle flows through the mitral valve into the left ventricle From the left ventricle most of the blood is pumped out through the aorta to the systemic circulation and is returned by the superior vena cava and the inferior vena cava to the right auricle A small volume of blood is forced through the ventricular septal defect into the diminutive right ventricle from which it is pumped into the pulmonary artery This is the only blood which reaches the lungs The blood which reaches the lungs is returned in the normal fashion to the left auricle There the cycle starts again

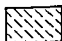
Clinical diagnosis is based on the contour of the heart In the anterior posterior positi [illegible]
The [illegible]
to th [illegible]
the s[illegible] of blood which reaches the lungs for oxygenation cyanosis is intense After the closure of the ductus arteriosus the blood can still reach the lungs by way of the pulmonary artery therefore, the condition is compatible with life for a longer time than when tricuspid atresia is combined with a non functioning right ventricle The electrocardiogram shows a left axis deviation and evidence of left ventricular preponderance

Diagram VIII–3

Head and upper extremities
R Lung
L Lung
SVC
PA
PV
Pul. A
Pul V
Aorta
Pulmonary Artery
LA
RA
LV
RV
Descending Aorta
IVC
Infundibulum Stenosis
Defect in Interventricular Septum
Trunk and lower extremities

 Arterial blood (fully saturated)

 Venous and arterial blood
Cyanosis visible

 Small admixture of venous blood
No visible cyanosis

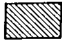
 Venous blood

Diagram VIII–3

Tricuspid atresia and a gross auricular septal defect combined with defective development of the right ventricle infundibular stenosis and a small ventricular septal defect

The essential features of this malformation are tricuspid atresia and defective development of the right ventricle Inasmuch as the tricuspid valve is atretic, the blood must flow from the right auricle to the left auricle it follows that either the foramen ovale is held open by the high pressure in the right auricle or there is a defect in the auricular septum The only way for the blood to enter the right ventricle is from the left ventricle through a ventricular septal defect There may also be infundibular stenosis of the right ventricle which places an additional obstruction to the flow of blood through the right ventricle to the pulmonary artery

The blood flows from the right auricle through the defect in the auricular septum into the left auricle where it meets the blood which is returned by the pulmonary veins to the left auricle This mixture of venous blood from the right auricle and oxygenated blood from the left auricle flows through the mitral valve into the left ventricle. From the left ventricle most of the blood is pumped out through the aorta to the systemic circulation and is returned by the superior vena cava and the inferior vena cava to the right auricle A small volume of blood is forced through the ventricular septal defect into the diminutive right ventricle from which it is pumped into the pulmonary artery This is the only blood which reaches the lungs The blood which reaches the lungs is returned in the normal fashion to the left auricle There the cycle starts again

Clinical diagnosis is based on the contour of the heart In the anterior posterior position the infundibular stenosis causes an absence of fullness of the pulmonary conus The small size of the right ventricle is seen in the left anterior-oblique position Owing to the complete admixture of the oxygenated and venous blood in the left auricle and the small volume of blood which reaches the lungs for oxygenation cyanosis is intense After the closure of the ductus arteriosus the blood can still reach the lungs by way of the pulmonary artery therefore, the condition is compatible with life for a longer time than when tricuspid atresia is combined with a non functioning right ventricle The electrocardiogram shows a left axis deviation and evidence of left ventricular preponderance

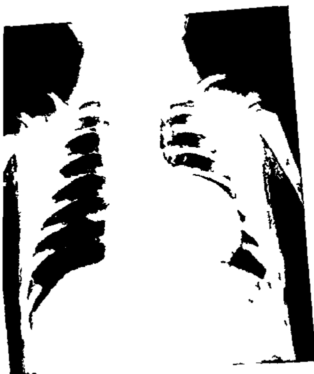
cles and a single ventricle If the opening in the auricular septum is relatively small, that is smaller than that of the normal tricuspid orifice, it is difficult for the right auricle to expel the blood The condition is functionally similar to that of an acquired tricuspid atresia in which it is difficult for the auricle to expel the blood through the stenosed tricuspid valve Both conditions are characterized by the same clinical phenomenon namely, the auricular pulsations are transmitted to the liver and cause presystolic pulsations at its margin

The differentiation of a presystolic pulsation from a systolic pulsation may be difficult, if not impossible, to determine, owing to the rapid rate of the infant s heart The two conditions can, however, be distinguished by the size of the right ventricle and not infrequently by the size of the liver The size of the right ventricle is determined by fluoroscopy and that of the liver by palpation A systolic pulsation occurs in association with a great dilatation of the right ventricle, tricuspid insufficiency, and engorgement of the liver In contrast to this, a presystolic pulsation in infancy is associated with a tricuspid atresia or marked hypoplasia, which in turn occurs in conjunction with a diminutive or an absent right ventricle and a liver of normal size It follows that in this malformation a pulsating liver of normal size combined with the absence of right ventricular enlargement is presumptive evidence of tricuspid atresia and a well formed auricular septum with a relatively small defect

A gross defect in the auricular septum, when sufficiently large to permit the free flow of blood from the right auricle to the left auricle, relieves the pressure in the right auricle, hence the liver does not pulsate

X RAY AND FLUOROSCOPIC FINDINGS

The absence of the right ventricle causes a characteristic change in the contour of the heart In the anterior posterior position the absence of the right ventricle is suggested by the absence of the shadow cast by the pulmonary conus of the right ventricle The upper margin of the cardiac shadow to the left of the sternum, instead of showing its usual convexity, is concave (Figures viii–6, 7, 8) In the left anterior-oblique position the diminutive size of the right ventricle is indicated by the fact that the cardiac shadow does not project toward the anterior chest wall beyond the shadow cast by the aorta (see Figures viii–6 and 7) The left ventricle, on the other hand, is enlarged and a considerable degree of rotation is required for the left ventricle to clear the spinal column Therefore, in spite of the absence of the right ventricle, the heart appears to be slightly enlarged, the enlargement, however, is entirely due to the increased size of the left ventricle

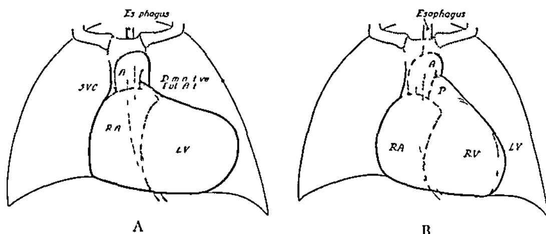


Anterior posterior position



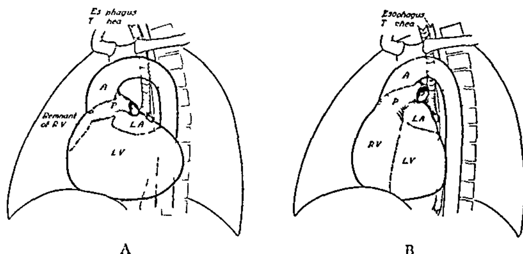
Left anterior-oblique position

Figure VIII-6 Tricuspid atresia, a non functioning right ventricle, and a well formed auricular septum Infant



A B

ANTERIOR POSTERIOR POSITION



A B

LEFT ANTERIOR-OBLIQUE POSITION

FIGURE VIII–7 (A) Tricuspid atresia and a non functioning right ventricle and (B) normal heart Infant

The diminutive size of the pulmonary artery causes the shadow cast by the great vessels in the anterior posterior position to be narrow Inasmuch as there is but a single vessel, upon rotation of the patient into the left anterior oblique position there is no increase in the width of this shadow In addition, the diminutive pulmonary artery renders the pulmonary window abnormally clear

Absence of the auricular septum results in a single auricle Usually the portion of the auricle which structurally represents the right auricle is more distensible and dilates more than the left side In early infancy the increased pressure on the right side of the auricle causes dilatation of the superior vena cava,

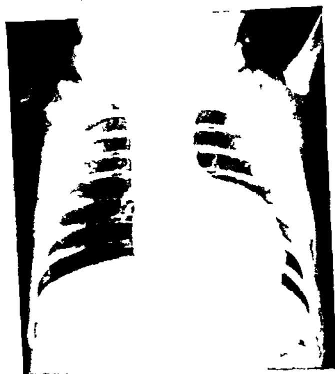


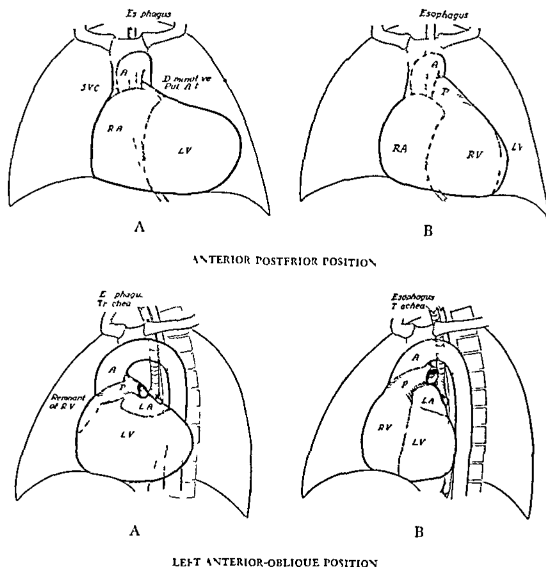
FIGURE VIII-8 Tricuspid atresia a non functioning right ventricle, and a gross auricular septal defect (same patient as in Figure VIII-1) Infant

hich is readily seen by x ray and fluoroscopy, as illustrated in Figures VIII-9 nd 10

This malformation may be compatible with life for a number of years When this occurs, it is common to find that the pulmonary artery arises from he outflow tract of the right ventricle in the normal manner and that the rudi nentary right ventricle receives its blood from the left ventricle through a defect n the ventricular septum

If the infant survives to childhood, as the patient grows the diaphragm de scends and the contour of the heart comes to resemble that of a tetralogy of Fallot Generally, however, the cardiac contour is more nearly square than in the tetralogy of Fallot This is due to the straight margin of the right auricle and to the slight fullness in the region of the pulmonary conus caused by the rudimentary outflow tract of the right ventricle and the relatively normal size of the pulmonary artery (see Figures VIII-11 and 12)

The hilar shadows may become accentuated as collateral circulation develops early and increases rapidly to compensate for the reduced flow through the main

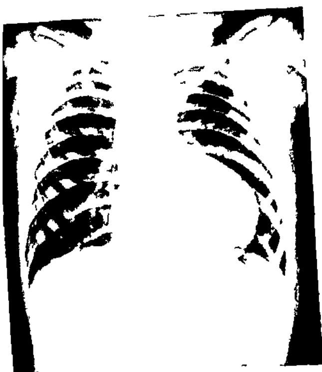


LEFT ANTERIOR-OBLIQUE POSITION

FIGURE VIII–7 (A) Tricuspid atresia and a non functioning right ventricle and (B) normal heart Infant

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Anterior posterior position



Left anterior oblique position

FIGURE VIII-11 Tricuspid atresia and a rudimentary right ventricle (same patient as in Figure VIII-2) Child



FIGURE VIII–9 Tricuspid atresia, a non functioning right ventricle, and a single auricle Infant

Note the dilatation of the superior vena cava

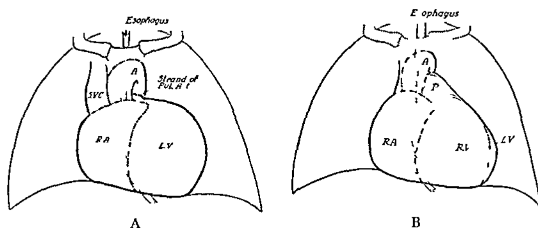
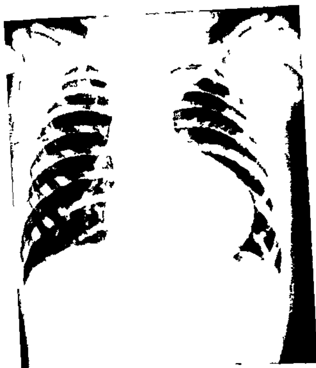


FIGURE VIII–10 (A) Tricuspid atresia, a non functioning right ventricle, a single auricle, and a dilated superior vena cava and (B) normal heart Infant

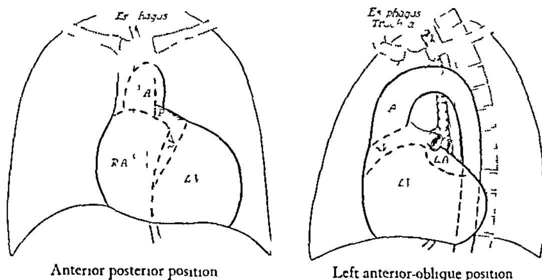


Anterior posterior position



eft anterior-oblique position

FIGURE VIII–11 Tricuspid atresia and a rudimentary right ventricle (same patient as in Figure VIII–2) Child



Anterior posterior position

Left anterior-oblique position

Figure viii–12 Tricuspid atresia Child

pulmonary artery These shadows are an aggregate of small shadows and do not pulsate The periphery of the lungs remains clear

In the left anterior oblique position, as the heart drops down, the anterior border of the heart and the aorta form an almost vertical line, which is frequently cut in toward the spinal column at the level of the diaphragm The aortic shadow is narrow and the pulmonary window is clear (see Figures viii–11 and 12)

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiographic findings are of diagnostic importance The finding of a left axis deviation in the standard leads, combined with evidence of left ventricular hypertrophy in the unipolar precordial leads, is always suggestive of tricuspid atresia Occasionally in young infants the standard leads may show a balanced axis, or in rare instances there may be even a right axis deviation The unipolar precordial leads, however, will uniformly show evidence of dominance of the left ventricle both in V_1 and in V_5 and V_6 (see Figure viii–13) this finding is present at birth [2]

When there is but a small defect in the auricular septum, the P waves may be excessively tall and peaked, they are frequently over 5 mm and occasionally 10 mm in height These are the so called Himalayan P waves When pulsations are palpable at the margin of the liver in a patient who has Himalayan P waves in the electrocardiogram, the defect in the auricle is certain to be small

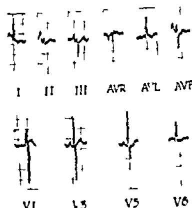


FIGURE VIII-15 Tricuspid atresia

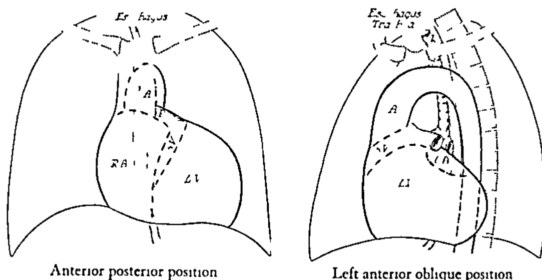
SPECIAL TESTS

The hematology is similar to that of other patients with persistent oxygen unsaturation of the arterial blood. The red blood cell count, the amount of available hemoglobin, and the hematocrit reading are all abnormally high. Over the years these patients also show a reduction in the number of blood platelets and in the blood fibrinogen.

The circulation time is usually shorter than normal as the blood passes directly from the right auricle to the left auricle and thence to the left ventricle and the systemic circulation.

The oxygen saturation of the arterial blood is greatly reduced and falls still further with exercise. The arterial oxygen saturation is lower when there is pulmonary atresia than when the pulmonary artery comes off the outflow tract of the right ventricle. Under the former circumstance closure of the ductus arteriosus is generally fatal. The determination of the oxygen saturation of the arterial blood is frequently the best guide in the evaluation of the severity of the condition. Although the author has known one patient who lived to thirty five years of age in whom the resting oxygen saturation of the arterial blood was only 35 per cent, such a saturation when obtained in an infant at rest is ominously low.

Cardiac catheterization generally gives little information concerning the presence of tricuspid atresia, as it is frequently impossible to pass the catheter beyond the right auricle. If, however, the diagnosis is wrong, the catheter will pass through the tricuspid valve into the right ventricle, thereby demonstrating that the tricuspid valve is not atretic.



Anterior posterior position Left anterior oblique position

FIGURE VIII-12 Tricuspid atresia Child

pulmonary artery These shadows are an aggregate of small shadows and do not pulsate The periphery of the lungs remains clear

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Figure VIII [illegible] Auricular septal defect with a rudimentary right ventricle and pulmonary stenosis Child

Anterior posterior and lateral films taken simultaneously

Angiocardiography is not of great diagnostic aid. The left auricle is, of course, visualized immediately after the right auricle. This finding does not, however, offer reliable evidence concerning the size of the auricular defect; it does not even aid in the differentiation of a gross defect in the auricular septum from patency of the valve covering the foramen ovale. Immediately after the left auricle is filled, the dye enters the left ventricle and thereafter both the pulmonary artery and the aorta are filled. Figure VIII–14 shows the dye in both the auricles. Figure VIII–15, which was taken a fraction of a second later, shows dye in the left ventricle and also in both the pulmonary artery and the aorta. The lateral view of Figure VIII–15 shows that the pulmonary artery lies anterior to the aorta.

DIAGNOSIS

The diagnosis is based primarily on the finding of persistent cyanosis and electrocardiographic evidence of left ventricular hypertrophy. This combination of findings always suggests a serious abnormality of the tricuspid valve and of the right ventricle. Murmurs are of no diagnostic significance. In infants, and occasionally in children, the contour of the heart may also be sufficiently char-

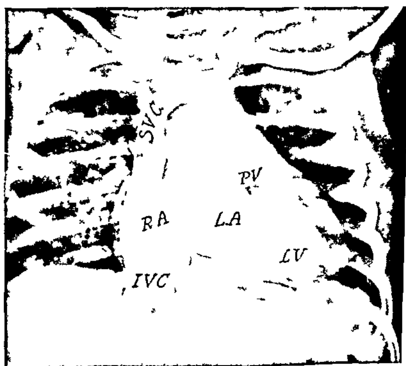


FIGURE VIII–14. Auricular septal defect with a rudimentary right ventricle and pulmonary stenosis. Child.

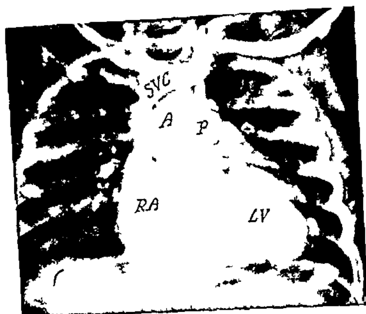




FIGURE VIII-15 Auricular septal defect with a rudimentary right ventricle and pulmonary stenosis Child

Anterior posterior and lateral films taken simultaneously

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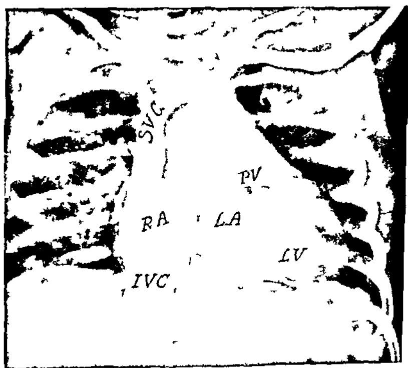


FIGURE VIII–14 Auricular septal defect with a rudimentary right ventricle and pulmonary stenosis Child

acteristic to suggest the diagnosis The significant x ray finding is the absence of the pulmonary conus in the anterior posterior position, combined with the absence of right ventricular enlargement in the left anterior-oblique position

DIFFERENTIAL DIAGNOSIS

In young infants the condition requires differentiation from a tetralogy of Fallot and from a single ventricle with pulmonary stenosis, and occasionally from a truncus arteriosus with markedly reduced pulmonary blood flow, and also from other malformations associated with defective development of the right ventricle, notably defective development of the right ventricle and pulmonary stenosis combined with an intact ventricular septum (see Chapter IX) and occasionally from tricuspid atresia combined with complete transposition of the great vessels with or without a single ventricle (see Section B)

A tetralogy of Fallot, especially in children, may closely resemble tricuspid atresia The clinical syndrome may be indistinguishable The two conditions are to be differentiated by the electrocardiographic findings In a tetralogy of Fallot there is usually a right axis deviation and evidence of right ventricular hypertrophy, whereas in a tricuspid atresia the electrocardiogram generally shows a left axis deviation and, regardless of age, almost invariably shows evidence of left ventricular dominance in V_1

A single ventricle with a rudimentary outlet chamber may require differentiation from a tricuspid atresia combined with defective development of the right ventricle In infancy the contours of the heart are similar in the two malformations when viewed in the left anterior-oblique position, but in the anterior posterior position their shapes are usually quite different In infancy a single ventricle with a rudimentary outlet chamber usually shows fullness of the pulmonary conus

As the infant grows to childhood and the diaphragm descends, the heart comes to occupy a more vertical position and the fullness caused by the rudimentary outflow chamber disappears The differential diagnosis is especially difficult when the electrocardiogram shows a left axis deviation, as in the case reported by Neill and Brink [2] The unipolar precordial leads are usually of aid in the differentiation of the two conditions In tricuspid atresia there is evidence from birth of left ventricular dominance in V_1, whereas in a single ventricle, although there may be a prominent R in V_5 or V_6, the left ventricular pattern is seldom pronounced in V_1

A single ventricle may occur in combination with pulmonary stenosis In

deed, the difference between a single ventricle and a rudimentary right ventricle which communicates with the left ventricle is primarily a matter of the formation of the ventricular septum In the former the outflow tract of the right ventricle appears as a pouch of the common ventricle, in the latter the septal wall is well formed and there is a relatively small defect through which the outflow tract communicates with the left ventricle (compare Figure VIII-2 with Figure XV-1) Cardiac catheterization will show that the common ventricle is entered directly from the right auricle

Truncus arteriosus with markedly reduced pulmonary blood flow may have a similar contour to the heart in the anterior posterior position, except that the aortic knob is usually more conspicuous In the left anterior-oblique position a truncus arteriosus shows enlargement of the right ventricle, this finding clearly distinguishes it from a non functioning right ventricle Further, the electrocardiogram shows evidence of hypertrophy of both ventricles and not a preponderant hypertrophy of the left ventricle Upon cardiac catheterization it will be possible to pass the catheter from the right auricle into the right ventricle and out into the aorta

Complete transposition of the great vessels may occur in combination with tricuspid atresia and defective development of the right ventricle (see Section B)

Defective development of the right ventricle and pulmonary stenosis with a normal tricuspid orifice and no over riding of the aorta is a rare malformation, nevertheless, it does occur (see Chapter IX) Occasionally this malformation is associated with a left axis deviation and evidence of left ventricular hypertrophy Usually, however, some evidence of right ventricular hypertrophy can be detected in V_1 or V_{3R} These findings give the clue to the diagnosis Cardiac catheterization will usually show high pressure in the right ventricle Angiocardiography will show that a small pulmonary artery arises from the right ventricle, but the aorta will not be simultaneously delineated

TREATMENT

In patients with tricuspid atresia combined with pulmonary stenosis or atresia the primary difficulty is lack of adequate circulation to the lungs Consequently the e patients may be greatly helped by a Blalock Taussig operation or by a Potts anastomosis

There is, however, always the danger that the increased volume of blood returned from the lungs will raise the pressure in the left auricle to such an extent that the right auricle will have difficulty in expelling its blood to the left auricle

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A single ventricle may occur in combination with pulmonary stenosis In

have become sufficiently enlarged to be of functional importance in the maintenance of the pulmonary circulation, or the pulmonary artery persists as a functioning tube which arises from a small right ventricle, which in turn receives its blood from the left ventricle Under such circumstances the condition may be compatible with life for a number of years Nevertheless, most of these patients develop severe polycythemia and are extremely incapacitated A Blalock Taussig operation may greatly improve the prognosis

SUMMARY

Tricuspid atresia is always associated with defective development of the right ventricle Usually this chamber is absent or exists as a diminutive non functioning chamber In addition, there is generally pulmonary atresia Occasionally, when there is tricuspid atresia, the primitive outflow tract develops into a rudimentary outflow chamber which receives blood from the left ventricle Under such circumstances the pulmonary artery may arise in the normal fashion Nevertheless, there is difficulty in the direction of blood to the lungs, hence there is real or functional pulmonary stenosis except in the rare instances in which there is a complete transposition of the great vessels (see Section B)

When the tricuspid valve is atretic the blood from the right auricle is shunted into the left auricle and thence to the left ventricle The mixture of venous and arterial blood reaches the lungs either through a stenosed pulmonary orifice or by way of the ductus arteriosus Consequently there is both a mixture of venous and arterial blood in the left auricle and a reduction in the pulmonary blood flow Hence there is persistent cyanosis

Difficulty in feeding and failure to gain are common complaints

Attacks of paroxysmal dyspnea are common and may be fatal

Squatting is common in patients who survive to childhood

The liver may or may not be enlarged and there may or may not be pulsations palpable at the margin of the liver The occurrence of a pulsating liver which is not engorged is indicative of tricuspid atresia and only a small opening in the auricular septum

The heart is but slightly enlarged The second sound at the base is pure Murmurs may or may not be present.

The diagnosis is suggested by the shape of the heart as seen in the x ray or by fluoroscopy In the anterior posterior view the shadow cast by the pulmonary conus is absent In the left anterior-oblique position the right ventricle does not project forward toward the anterior chest wall beyond the margin of the aorta and the pulmonary window is abnormally clear As the patient grows, the heart

This is notably true when there is only a small defect in the auricular septum or when the foramen ovale is covered by a valve Under such circumstances the increased volume of blood returned from the lungs will close the foramen ovale and the patient will develop severe right sided heart failure Therefore, if the defect in the auricular septum is small, it may be necessary to enlarge the auricular defect as well as to perform the Blalock-Taussig operation

Although some patients have been greatly helped by the combined procedure, over a period of years results have not been as good as those for patients with a tetralogy of Fallot, some patients with tricuspid atresia develop progressive cardiac enlargement and die of cardiac failure within six to eight months after operation Inasmuch as the tricuspid orifice is atretic and the right ventricle is abnormally small, there is virtually no hope of corrective surgery

Glenn[3] has proposed an end to-end anastomosis of the superior vena cava to the right pulmonary artery in order to direct venous blood to the lungs and by pass the right ventricle The operation has proved beneficial to some patients but carries the risk of cerebral edema The long time results cannot as yet be evaluated but decrease in the size of the anastomosis would increase the cerebral pressure

In infants failure to gain or the occurrence of attacks of paroxysmal dyspnea is usually an indication for operation Whenever possible it is advisable to determine the oxygen saturation of the arterial blood If the oxygen saturation of the arterial blood at rest is 20 per cent or less, the danger of death from anoxemia is so great that it is justifiable to assume the risk involved in operation

In older children, however, it is important to remember that after operation there is far greater danger of development of cardiac failure for these patients than for patients with a tetralogy of Fallot Therefore, only if the child is severely incapacitated is operation justified Nevertheless, some children have been greatly helped for many years One of the author s patients did well for eight years and then cyanosis recurred, now he has again been improved by a second anastomosis performed ten years after his first operation

PROGNOSIS

When tricuspid atresia is combined with a non functioning right ventricle and pulmonary atresia, the prognosis is extremely poor As the ductus arteriosus undergoes obliteration the condition usually becomes incompatible with life, the duration of life is usually but three to four months Infants, however, have been known to live for fifteen months When a patient survives for more than two years, either the ductus arteriosus has remained patent, the bronchial arteries

circumstances the right ventricle usually develops as a small chamber which receives its blood from the left ventricle The aorta arises from the right ventricle and the pulmonary artery from the left ventricle Inasmuch as the right ventricle lacks its normal inflow tract, some opening in the auricular septum is inevitable, either the foramen ovale remains patent or there is a gross defect in the auricular septum Furthermore, since *no blood enters the right ventricle from the right auricle*, the only way for blood to reach the right ventricle is from the left ventricle, there must also be a defect in the ventricular septum *Indeed, if there is no* defect in the ventricular septum, the right ventricle is a blind sac and the transposed aorta is markedly hypoplastic and atretic at its base

When the defect in the ventricular septum is small, the aorta, which arises from the right ventricle, may be hypoplastic, as shown in Figure VIII–16 When, however, the defect in the ventricular septum is relatively large, the aorta may be of normal size

The pulmonary artery also may vary in size, it may be of normal size with or without pulmonary stenosis, or it may be abnormally small Figure VIII–17 shows a heart in which there was tricuspid atresia, a relatively large defect in the ventricular septum with an aorta of normal size arising from the right ventricle, and an abnormally small pulmonary artery arising from the left ventricle

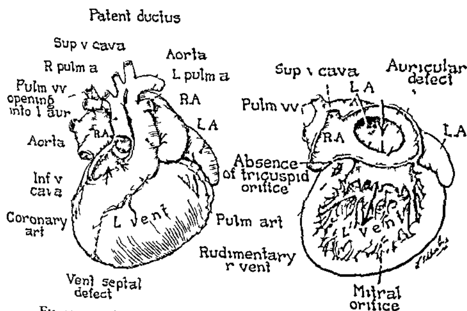


FIGURE VIII 16 Tricuspid atresia combined with complete transposition of the great vessels and a hypoplastic aorta arising from a rudimentary right ventricle Infant

comes to occupy a more vertical position and its contour becomes similar to that of a tetralogy of Fallot In the anterior posterior view the contour is, however, somewhat square and in the left anterior oblique position the anterior border of the heart and the aorta form an almost vertical line

The electrocardiogram usually shows a left axis deviation and the unipolar precordial leads uniformly show evidence of left ventricular dominance in V_1 This finding is so constant that its presence should always suggest a serious abnormality of the tricuspid valve and of the right ventricle

Laboratory studies show evidence of polycythemia The circulation time is abnormally short The oxygen saturation of the arterial blood is reduced and falls still further with exercise Cardiac catheterization gives little information, as it is only possible to pass the catheter from the right auricle to the left auricle Angiocardiography is not of great diagnostic aid

The diagnosis is based on the clinical, fluoroscopic, and the all important electrocardiographic findings

The malformation requires differentiation from a tetralogy of Fallot and from a single ventricle with pulmonary stenosis, and occasionally from a truncus arteriosus with markedly reduced pulmonary blood flow and also from defective development of the right ventricle without tricuspid atresia

Treatment is directed to increase the pulmonary blood flow, and either a Blalock anastomosis or a Potts procedure will help If the defect in the auricular septum is small, it is necessary to combine a systemic pulmonary anastomosis with enlargement of the defect in the auricular septum The long time results are not as good as for a patient with a tetralogy of Fallot, because after operation a number of patients develop progressive cardiac enlargement and intractable cardiac failure Glenn s operation may be of real benefit.

The prognosis in patients with a non functioning right ventricle is extremely poor If the right ventricle receives some blood from the left ventricle, the patient may survive for a number of years Such patients are usually severely incapacitated The prognosis may be greatly improved by operation

B *Tricuspid Atresia Combined with Complete Transposition of the Great Vessels*

NATURE OF THE MALFORMATION

The essential feature of the malformation is the combination of a complete transposition of the great vessels and atresia of the tricuspid valve Under such

circumstances the right ventricle usually develops as a small chamber which receives its blood from the left ventricle The aorta arises from the right ventricle and the pulmonary artery from the left ventricle Inasmuch as the right ventricle lacks its normal inflow tract, some opening in the auricular septum is inevitable, either the foramen ovale remains patent or there is a gross defect in the auricular septum Furthermore, since no blood enters the right ventricle from the right auricle, the only way for blood to reach the right ventricle is from the left ventricle, there must also be a defect in the ventricular septum Indeed, if there is no defect in the ventricular septum, the right ventricle is a blind sac and the transposed aorta is markedly hypoplastic and atretic at its base

When the defect in the ventricular septum is small, the aorta, which arises from the right ventricle, may be hypoplastic, as shown in Figure VIII–16 When, however, the defect in the ventricular septum is relatively large, the aorta may be of normal size

The pulmonary artery also may vary in size, it may be of normal size with or without pulmonary stenosis, or it may be abnormally small Figure VIII–17 shows a heart in which there was tricuspid atresia, a relatively large defect in the ventricular septum with an aorta of normal size arising from the right ventricle, and an abnormally small pulmonary artery arising from the left ventricle

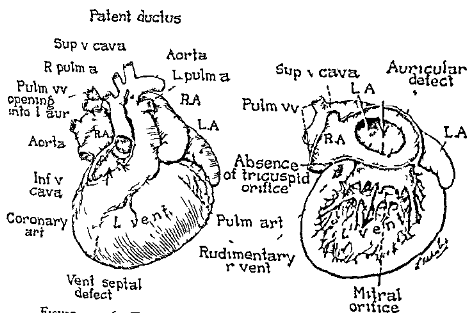


FIGURE VIII–16 Tricuspid atresia combined with complete transposition of the great vessels and a hypoplastic aorta arising from a rudimentary right ventricle Infant

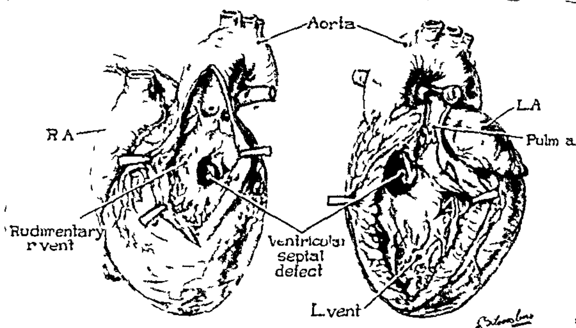
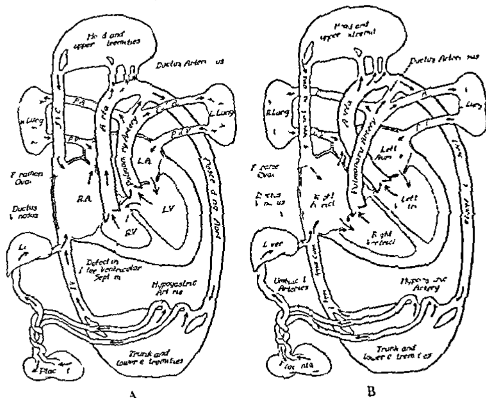


FIGURE VIII-17 Tricuspid atresia combined with complete transposition of the great vessels and a small pulmonary artery arising from the left ventricle (same patient as in Figure VIII-19) Child

COURSE OF THE CIRCULATION

During fetal life the circulation is altered both by the absence of the tricuspid orifice and by the abnormal position of the aorta All the blood from the right auricle must flow to the left auricle and thence to the left ventricle If there is aortic atresia, all the blood from the left ventricle is pumped out through the pulmonary artery to the lungs and through the ductus arteriosus to the body When, however, the aorta comes off the right ventricle, some blood is pumped out through the pulmonary artery to the lungs and some through the septal defect into the right ventricle and thence into the aorta The blood which flows to the lungs is returned in the normal manner to the left auricle and that which flows to the body is returned by the superior vena cava and inferior vena cava to the right auricle, as shown in Figure VIII-18

After birth the course of the circulation remains essentially the same The atresia of the tricuspid orifice causes all the blood from the right auricle to flow into the left auricle, where it meets the blood returned from the lungs This mixture of oxygenated and venous blood flows into the left ventricle If there is aortic atresia, all the blood from the left ventricle is pumped out into the pulmonary artery Thence part of the blood flows to the lungs and part through the



A B

FIGURE VIII-18 Fetal circulation (A) Tricuspid atresia and defective development of the right ventricle combined with complete transposition of the great vessels and a high ventricular septal defect and (B) normal heart

ductus arteriosus to the aorta. The course of the circulation is shown in Diagram VIII-4 When the aorta arises from the right ventricle and receives its blood from the left ventricle, part of the blood from the left ventricle is pumped into the pulmonary artery and part is pumped through the ventricular septal defect to the right ventricle and thence into the aorta The blood which is pumped into the pulmonary artery goes to the lungs, where it is oxygenated, and returned by the pulmonary veins to the left auricle, the blood which is pumped from the right ventricle into the aorta goes to the body and is returned by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again (see Diagram VIII-5)

PHYSIOLOGY OF THE MALFORMATION

In this type of transposition the left auricle receives an admixture of venous blood from the right auricle and oxygenated blood from the lungs, this mixture

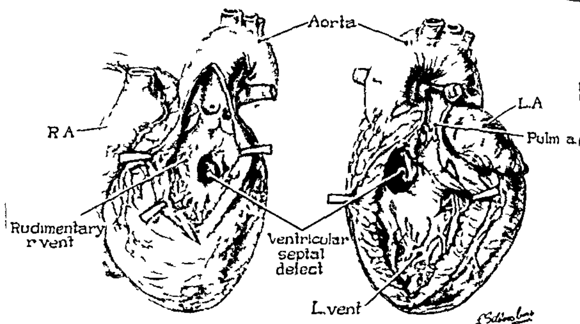


FIGURE VIII–17 Tricuspid atresia combined with complete transposition of the great vessels and a small pulmonary artery arising from the left ventricle (same patient as in Figure VIII–19) Child

COURSE OF THE CIRCULATION

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DIAGRAM VIII-4

Tricuspid atresia and a non functioning right ventricle combined with complete transposition of the great vessels an auricular septal defect atresia of the aorta at its base and a patent ductus arteriosus

This malformation is a combination of two anomalies namely, (1) a non functioning right ventricle and (2) a complete transposition of the great vessels combined with atresia of the aorta at its base

Owing to the tricuspid atresia the blood which enters the right auricle cannot leave by its normal pathway and must escape through a defect in the auricular septum into the left auricle In the left auricle there is complete admixture of the venous blood from the systemic circulation with the oxygenated blood returned by the lungs to the left auricle This mixture of oxygenated and venous blood passes into the left ventricle and is pumped out through the transposed pulmonary artery to the lungs Inasmuch as the aorta is atretic at its base the only way for the blood to reach the systemic circulation is through the ductus arteriosus The blood which is directed to the body is returned in the normal manner by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again

Clinical diagnosis Cyanosis is always intense The circulation to the body is poor Inasmuch as the blood is pumped from the left ventricle to the lungs and through the ductus arteriosus to the body there is systemic pressure in the pulmonary artery and hence there is pulmonary hypertension Moreover a large volume of blood is pumped to the lungs pulmonary congestion occurs early and may be extreme The circulation may be so poor that the duration of life is too short to produce cardiac hypertrophy Th heart is normal in size The x ray shows an absence of the fullness of the pulmonary conus in the anterior posterior position and a small right ventricle in the left anterior-oblique position The electrocardiogram shows a left axis deviation and evidence of combined hypertrophy and left ventricular dominance

Diagram viii–4

Head and upper extremities
Ductus Arteriosus
R Lung
L Lung
SVC
PA
Pul A
Aorta
Pulmonary Artery
PV
Pul V
RA
LA
LV
RV
Clot
IVC
Descending Aorta
Trunk and lower extremities



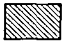
Arterial blood (fully saturated)



Venous and arterial blood
Cyanosis visible



Small admixture of venous blood
No visible cyanosis



Venous blood

Diagram VIII-5

Tricuspid atresia and complete transposition of the great vessels an auricular septal defect and a ventricular septal defect

This malformation combines a tricuspid atresia and a complete transposition of the great vessels The occurrence of the tricuspid atresia renders inevitable a defect in the auricular septum The origin of the aorta from the right ventricle which lacks its normal inflow tract, renders inevitable a defect in the ventricular septum

Because of the tricuspid atresia the blood from the right auricle cannot escape by way of its normal channel it can escape only through the defect in the auricular septum into the left auricle. There it mixes with the blood which enters the left auricle from the pulmonary veins This admixture of venous and arterial blood flows into the left ventricle Part of the blood from the left ventricle is pumped out by way of the pulmonary artery to the lungs and the oxygenated blood is again returned to the left auricle and the left ventricle Part of the blood is forced through the ventricular septal defect into the right ventricle and thence it is pumped out by way of the aorta to the systemic circulation. The blood from the head and the upper extremities is returned in the normal fashion by the superior vena cava and that from the lower extremities is returned by the inferior vena cava to the right auricle. There the cycle starts again

Clinical diagnosis The complete admixture of venous and arterial blood, combined with the difficulty in the direction of the blood to the systemic circulation causes intense cyanosis The tricuspid atresia causes the right ventricle to be smaller than the left ventricle. Consequently the electrocardiogram shows a left axis deviation and evidence of combined hypertrophy and left ventricular preponderance The complete transposition of the great vessels leads to progressive cardiac enlargement and cardiac failure The fact that the pulmonary artery is of normal size and arises from the left ventricle means that the lungs receive blood under systemic pressure The pulmonary circulation however is adequate or excessive and congestion of the lungs is common In brief, this malformation shows a combination of the features characteristic of a complete transposition of the great vessels and those which result from the tricuspid atresia and defective development of the right ventricle

DIAGRAM VIII–5

Head and upper extremities
R Lung
L Lung
S.V.C.
PA
PV
Aorta
pulmonary art.
Pul. A.
Pul V
L.A.
R.A.
L.V.
R.V.
Descending Aorta
I.V.C.
Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

Diagram VIII–5

Tricuspid atresia and complete transposition of the great vessels an auricular septal defect and a ventricular septal defect

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Clinical diagnosis The complete admixture of venous and arterial blood, combined with the difficulty in the direction of the blood to the systemic circulation, causes intense cyanosis The tricuspid atresia causes the right ventricle to be smaller than the left ventricle Consequently the electrocardiogram shows a left axis deviation and evidence of combined hypertrophy and left ventricular preponderance The complete transposition of the great vessels leads to progressive cardiac enlargement and cardiac failure The fact that the pulmonary artery is of normal size and arises from the left ventricle means that the lungs receive blood under systemic pressure The pulmonary circulation however is adequate or excessive and congestion of the lungs is common In brief this malformation shows a combination of the features characteristic of a complete transposition of the great vessels and those which result from the tricuspid atresia and defective development of the right ventricle

of venous and arterial blood flows into the left ventricle Consequently the same admixture of venous and arterial blood is pumped into the aorta and out through the pulmonary artery to the lungs If there is no stenosis of the pulmonary orifice, the total circulation to the lungs is normal or excessive but the effective flow is reduced Furthermore, the origin of the pulmonary artery from the left ventricle means that blood is pumped into the pulmonary artery under the same pressure as it is pumped into the aorta Consequently, unless there is stenosis of the pulmonary valve, there is always pulmonary hypertension The adequacy of the systemic circulation depends upon the size of the ventricular defect When the defect is minute or the aorta is atretic at its base, the condition is physiologically and clinically similar to that of aortic atresia and the duration of life is extremely short (see Chapter XIII, Section A)

The larger the defect the more readily is blood directed to the aorta hence the duration of life is longer Nevertheless, the condition usually leads to progressive cardiac enlargement and cardiac failure at an early age

If there is pulmonary stenosis, more blood is forced through the septal defect to the right ventricle, the systemic circulation is thereby increased and less blood goes to the lungs for oxygenation Under such circumstances, although the lungs are protected from the high pressure in the left ventricle, the effective flow may be greatly reduced and the oxygen saturation of the arterial blood is proportionally low

CLINICAL FINDINGS

Cyanosis dates from birth and is of uniform distribution It is, however, not necessarily intense

Respirations are rapid When there is pulmonary atresia or extreme pulmonary stenosis, the infant may suffer from attacks of paroxysmal dyspnea Such attacks are absent if the pulmonary artery is small but the pulmonary pressure is high

Pulmonary congestion occurs when the pulmonary artery is of normal size If the aorta is atretic, pulmonary congestion occurs early and remains severe throughout the infant s brief life The occurrence of pulmonary stenosis or an abnormally small pulmonary artery protects the lungs and renders the condition more compatible with life

The exercise tolerance of the child varies but it is not severely limited unless there is real difficulty in the direction of blood to the lungs

The liver is usually enlarged and may extend to the umbilicus

Edema is a late manifestation

CARDIAC FINDINGS

The size of the heart varies with the size of the aorta and the pulmonary artery When either of the great vessels is atretic, the duration of life is so short that the heart remains small

When both great vessels are of functional importance, the duration of life is longer Indeed, when the aorta is of normal size and the pulmonary artery is small or moderately stenosed, the condition may be compatible with life for a number of years The heart becomes moderately enlarged

The second heart sound at the base is accentuated as the aorta is displaced anteriorly and to the left Reduplication of the second heart sound indicates that both great vessels are of relatively normal size *Murmurs* are of no great diagnostic aid Usually there is a *systolic* murmur A *gallop rhythm* may or may not be present

Progressive cardiac enlargement is the rule when both great vessels are of normal size Consequently the condition leads to cardiac failure at an early age

Cardiac failure is manifested by congestion of the lungs, engorgement of the liver, and edema of the extremities There may even be ascites

X RAY AND FLUOROSCOPIC FINDINGS

The contour of the heart varies with the relative size of the great vessels, the age of the patient, and the duration of life

When the pulmonary artery is atretic the lungs are excessively clear and the contour of the heart is similar to that in other cases of a non functioning right ventricle, as shown in Figures VIII–6 and 8

When the aorta is atretic at its base and the pulmonary artery is of normal size, although the contour of the heart is similar to that which occurs with pulmonary atresia and a non functioning right ventricle, the lungs are not unduly clear but duration of life may be too short for hilar shadows to become conspicuous

When the aorta is of normal size and the pulmonary artery is abnormally small the contour of the heart is somewhat square This contour is accentuated when the aorta arches to the right and displaces the superior vena cava to the right (as shown in Figure VIII–19) and under such circumstances examination in the left anterior-oblique position will show that the right ventricle is small The lungs are clear The pulmonary window is also clear because the posteriorly placed pulmonary artery courses to the lungs at an abnormally low level The structure of the heart in this combination of anomalies is illustrated in Figure VIII–17 and the x ray of the case is shown in Figure VIII–19

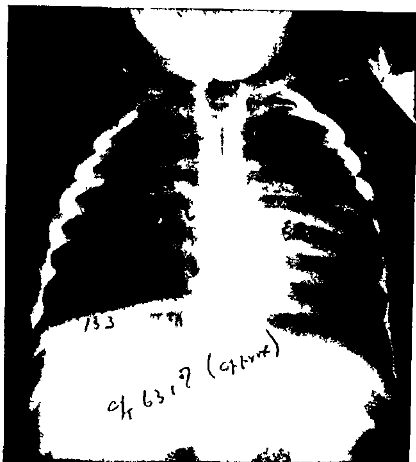


FIGURE viii–19 Tricuspid atresia combined with complete transposition of the great vessels and a small pulmonary artery (same patient as in Figure viii–17) Child

When both great vessels are of normal size, the heart undergoes progressive cardiac enlargement, as it does in other cases of complete transposition of the great vessels, both ventricles become enlarged and the right auricle may be dilated There is a concave curve at the base of the heart to the left of the sternum and congestion in the lungs, as shown in Figure viii–20 Examination of the patient in the left anterior oblique position shows great enlargement posteriorly and little or no enlargement anteriorly

ELECTROCARDIOGRAPHIC FINDINGS

The standard leads usually show a left axis deviation and almost invariably the unipolar precordial leads will show evidence of left ventricular dominance, which increases as the patient grows older

SPECIAL TESTS

The red blood cell count, the amount of available hemoglobin and *the hematocrit reading* are always increased

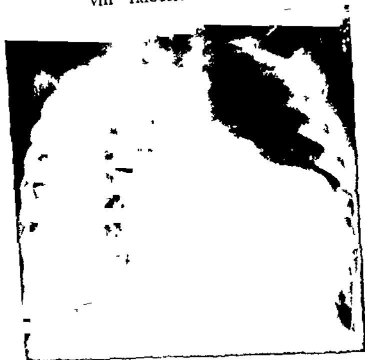


FIGURE VIII–20 Tricuspid atresia combined with complete transposition of the great vessels and a large pulmonary artery Child

The oxygen saturation of the arterial blood is always reduced

Cardiac catheterization is usually of no great diagnostic aid, as it is frequently impossible to pass the catheter beyond the right auricle

Angiocardiography shows prompt opacification of the left auricle immediately after the right auricle is filled, and before either ventricle is delineated When both great vessels are present, they are visualized simultaneously In the lateral film the aorta is seen to lie anterior to the pulmonary artery

DIAGNOSIS

The diagnosis is based upon the finding of a left axis deviation and evidence of left ventricular dominance in the electrocardiogram of a cyanotic patient with a heart which has a concave curve at its base to the left of the sternum When this finding is combined with increased vascularity of the lungs, it is clear evidence that the pulmonary artery is transposed and is of normal size

When the pulmonary artery is small the diagnosis is difficult, as the condition simulates a non functioning right ventricle without transposition of the great vessels The absence of paroxysmal dyspnea and of squatting suggests that the tricuspid atresia is combined with a transposition of the great vessels

When a complete transposition of the great vessels combined with atresia of

the aorta at its base is associated with tricuspid atresia and a non functioning right ventricle, the course of the circulation is similar to that of other types of aortic atresia and the contour of the heart and the electrocardiographic findings are similar to those of a non functioning right ventricle

DIFFERENTIAL DIAGNOSIS

The condition requires differentiation from tricuspid atresia without transposition of the great vessels, from a single ventricle, and from other types of complete transposition of the great vessels

Tricuspid atresia without transposition of the great vessels is always associated with reduced pulmonary blood flow As previously mentioned, the contour of the heart in this malformation closely resembles that which is found in tricuspid atresia when the great vessels are transposed Angiocardiography may be necessary to determine which of the great vessels lies anterior to the other

A single ventricle may be confused with tricuspid atresia and transposition of the great vessels Usually the electrocardiogram differentiates the two conditions If the electrocardiogram shows a left axis deviation and evidence of left ventricular hypertrophy, cardiac catheterization may be necessary to determine whether the left auricle or the common ventricle is entered from the right auricle

Other types of complete transposition of the great vessels show electrocardiographic evidence of a right axis deviation and right ventricular hypertrophy

TREATMENT

Medical treatment is palliative, surgical treatment is difficult and often impossible

When either of the great vessels is atretic the condition of the infant is too precarious for surgery to be attempted

When both great vessels are of functional importance there are three major considerations to be taken into account (1) the adequacy of the pulmonary blood flow, (2) the size of the opening between the two auricles, and (3) the size of the ventricular defect

When there is pulmonary stenosis, the circulation to the lungs may be increased by a systemic pulmonary anastomosis, but if such is attempted due consideration must be given to the size of the auricular defect If the only communication between the two auricles is the patency of the foramen ovale, the increased circulation to the lungs will close the valve and precipitate right sided

cardiac failure, therefore it may also be necessary to create an auricular defect The heart, however, may not be able to adjust to the altered circulation

In some instances the ventricular septal defect is so small that it is difficult for the left ventricle to expel the blood which it receives from the lungs The creation of a ventricular septal defect is far more difficult than that of an auricu lar defect At best it would only change the malformation to that of a single ventricle

PROGNOSIS

The prognosis is poor The duration of life depends upon the size of the great vessels and the size of the intracardiac shunts The condition frequently leads to progressive cardiac enlargement and cardiac failure Most infants with this mal formation die in the first year of life, a few may live to childhood

SUMMARY

Tricuspid atresia may occur in combination with a complete transposition of the great vessels Under such circumstances the right ventricle, from which the aorta arises, is of functional importance and receives its blood from the left ven tricle through a defect in the ventricular septum The pulmonary artery, which arises from the left ventricle, may be normal in size, abnormally large, or ste notic Unless there is pulmonary stenosis, there is always pulmonary hyperten sion Inasmuch as the aorta arises from the right ventricle, which receives its blood from the left ventricle, there may be difficulty in the establishment of the systemic circulation

Cyanosis dates from birth and is usually intense

The exercise tolerance of patients with this condition varies with the severity of the pulmonary stenosis and with the adequacy of the systemic circulation

The heart tends to undergo progressive enlargement The second sound over the pulmonary area is accentuated, murmurs are variable The x ray shows a somewhat square contour with a concave curve at the base of the heart to the left of the sternum The vascularity of the lung fields varies with the size of the pulmonary orifice

The condition requires differentiation from a single ventricle combined with a transposition of the great vessels, and from other types of tricuspid atresia and also other types of complete transposition of the great vessels

Treatment is usually unsatisfactory The structure of the heart is so grossly abnormal that there is little chance of surgical relief Only if the patient suffers

from pulmonary stenosis and lack of adequate pulmonary blood flow is there any hope that a systemic pulmonary anastomosis will help, and even then it may be necessary to combine this procedure with the creation of an auricular septal defect

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3 Glenn W W L Circulatory by pass of the right side of the heart iv Shunt between superior vena cava and distal right pulmonary artery report of clinical application New England J of Med 259 117-120, 1958

CHAPTER IX

DEFECTIVE DEVELOPMENT OF THE RIGHT VENTRICLE

DEFECTIVE development of the right ventricle may occur as an isolated malformation with an intact ventricular septum and without tricuspid atresia There are at least two types of abnormality in the development of the right ventricle associated with a normal tricuspid orifice The more common type is the one in which the right ventricle is abnormally small but is of functional importance in the direction of blood to the lungs In such instances the pulmonary artery is abnormally small and usually its orifice is stenotic In rare instances the pulmonary stenosis is so extreme that there is virtual pulmonary atresia The malformation differs from the usual pulmonary stenosis with an intact ventricular septum in that the right ventricle is so small that it cannot pump its full load and is unable to dilate to meet the demands of the body Clinically the condition more closely resembles a tetralogy of Fallot with severe pulmonary stenosis or atresia than it does a pulmonary stenosis with an intact ventricular septum This anomaly is presented in Section A

The other type occurs when the defect in the development of the right ventricle is of such a nature that the right ventricle is a large chamber with a thin, flabby wall The wall is so poorly developed that the right ventricle is an ineffective pump The electrocardiogram usually shows a left axis deviation This condition is discussed in Section B

A *Defective Development of the Right Ventricle with an Intact Ventricular Septum*

NATURE OF THE MALFORMATION

In this malformation the right ventricle fails to expand and grow normally it remains a small chamber A small tricuspid valve opens into it in the normal manner The ventricular septum is intact and a small pulmonary artery arises from this chamber Usually there is a cap like stenosis of the pulmonary valve Figure IX–1 illustrates a heart of this type The stenosis may be so extreme that there is a functional pulmonary atresia Sometimes the infundibular tract is extremely narrow but leads to a diminutive pulmonary valve with tiny but nor

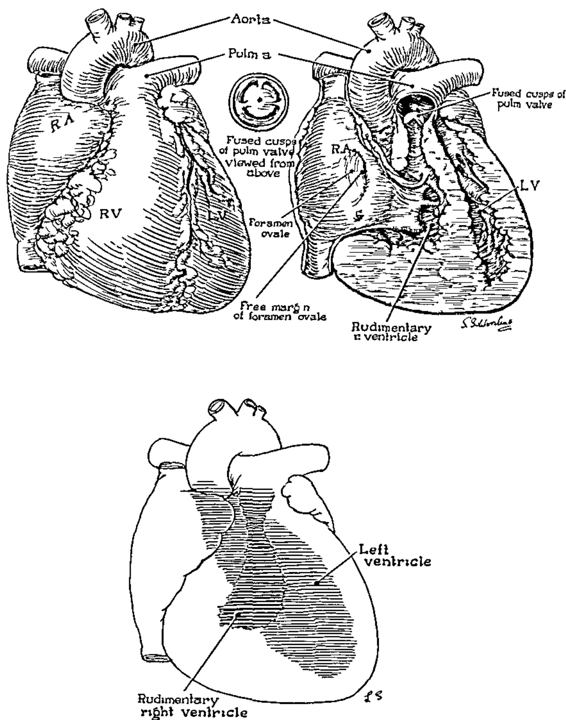


FIGURE IX-1 Defective development of the right ventricle with valvular pulmonary stenosis and an intact ventricular septum (Case IX-1) Child

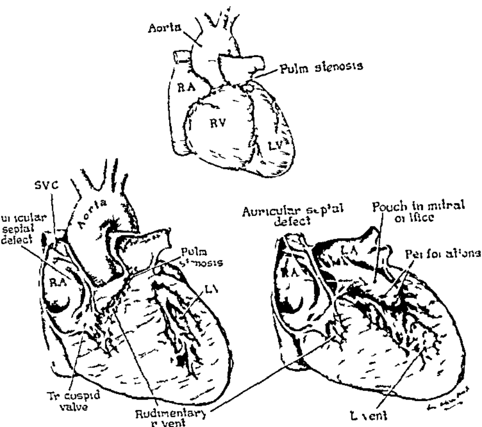


FIGURE IX–2 Defective development of the right ventricle with extreme infundibular stenosis

Note also the pouch extending from the left auricle into the left ventricle

mally formed cusps, as shown in Figure IX–2 In the latter instance the right ventricle was abnormally small, with marked narrowing of the outflow tract and a small pulmonary artery, and in addition there was an extraordinary membranous pouch which extended from the foramen ovale into the left auricle and partially obstructed the mitral orifice Consequently the infant suffered not only from an abnormally small right ventricle and pulmonary stenosis but also from mitral stenosis Little wonder that in this instance life was of such short duration that no studies were obtained

As in all types of malformations there are all grades of severity The right ventricle is always abnormally small but, if the pulmonary stenosis is not ex

tremely severe, the patient may live to childhood, whereas, if the stenosis is extremely severe or if there is pulmonary atresia, the condition leads to progressive cardiac enlargement and cardiac failure in early infancy If extreme pulmonary stenosis is combined with a diminutive right ventricle, the condition is functionally similar to a tricuspid atresia with a non functioning right ventricle

COURSE OF THE CIRCULATION

The blood from the right auricle is directed to the right ventricle in the normal manner and is pumped out through the stenosed pulmonary artery to the lungs, where it is oxygenated and returned in the normal manner to the left auricle The small size of the right ventricle renders it impossible for the right auricle to empty in the normal manner Hence the pressure in the right auricle is abnormally high, because of this the foramen ovale is held open and some blood is shunted from the right auricle into the left auricle There the venous blood from the right auricle meets the oxygenated blood returned from the lungs to the left auricle This admixture of venous and oxygenated blood flows into the left ventricle and is pumped out through the aorta to the systemic circulation and returned by the superior and inferior venae cavae to the right auricle There the cycle starts again, as shown in Diagram IX–I When there is pulmonary atresia, the course of the circulation is essentially the same as that shown in Diagram VIII–I

PHYSIOLOGY OF THE MALFORMATION

The small size of the right ventricle and the pulmonary obstruction increase the pressure in the right ventricle, which in turn raises the pressure in the right auricle The high pressure in the right auricle holds the foramen ovale open and a right to-left shunt is established at the auricular level The pulmonary stenosis protects the lungs, so that the pulmonary vascular bed is normal

CLINICAL FINDINGS

Cyanosis is usually present at birth or shortly thereafter and increases as the ductus arteriosus undergoes obliteration

Clubbing of the extremities occurs early

Polycythemia develops at an early age The red blood cell count, the amount of available hemoglobin, and the hematocrit reading are all abnormally high If the duration of life permits, polycythemia may become extreme

Dyspnea is common but attacks of paroxysmal dyspnea rarely occur

Growth and *development* are retarded Infants with this malformation usually gain weight extremely slowly

Exercise tolerance is limited but the child seldom squats when tired When short of breath, he simply stops to rest As the child grows, he gradually becomes progressively more incapacitated This is in striking contrast to the child with a tetralogy of Fallot, who usually improves between two and five years of age

The liver is often enlarged and may be huge, frequently there are *pulsations* palpable at the margin of the liver

Edema and *ascites* are late manifestations

CARDIAC FINDINGS

The size of the heart varies with the severity of the abnormality The heart is always somewhat enlarged The condition leads to progressive cardiac enlargement The rate of enlargement varies with the size of the right ventricle and the severity of the pulmonary stenosis Thus there may be only slight enlargement at six years of age, as shown in Figure IX-3, or there may be tremendous enlargement at four months of age, as shown in Figure IX-4 The *second sound* to the left of the sternum is weak or absent A *systolic murmur* may or may not be audible

The malformation causes great cardiac enlargement and terminal cardiac failure with engorgement of the liver and edema of the extremities and even ascites The lungs, however, usually remain clear

X RAY AND FLUOROSCOPIC FINDINGS

The heart is usually enlarged and may be huge In the anterior posterior position there is a narrow pedicle at the base of the heart and the upper margin of the shadow to the left of the sternum has a concave curve The right auricle is slightly enlarged and the vascular markings are decreased, as shown in Figure IX-3

In the left anterior-oblique position there is no enlargement of the right ventricle but the left ventricle extends abnormally far posteriorly Owing to the pulmonary stenosis the pulmonary window is clear

If the pulmonary stenosis is extreme, the right auricle and the left ventricle are enlarged, the heart lies horizontally upon the diaphragm, and the lungs are excessively clear as shown in Figure IX-4 Under such circumstances, in the left anterior-oblique view, the enlargement of the right auricle may extend so far anteriorly as to suggest great right ventricular enlargement and the ventricular

tremely severe, the patient may live to childhood, whereas, if the stenosis is extremely severe or if there is pulmonary atresia, the condition leads to progressive cardiac enlargement and cardiac failure in early infancy. If extreme pulmonary stenosis is combined with a diminutive right ventricle, the condition is functionally similar to a tricuspid atresia with a non-functioning right ventricle.

COURSE OF THE CIRCULATION

The blood from the right auricle is directed to the right ventricle in the normal manner and is pumped out through the stenosed pulmonary artery to the lungs, where it is oxygenated and returned in the normal manner to the left auricle. The small size of the right ventricle renders it impossible for the right auricle to empty in the normal manner. Hence the pressure in the right auricle is abnormally high; because of this the foramen ovale is held open and some blood is shunted from the right auricle into the left auricle. There the venous blood from the right auricle meets the oxygenated blood returned from the lungs to the left auricle. This admixture of venous and oxygenated blood flows into the left ventricle and is pumped out through the aorta to the systemic circulation and returned by the superior and inferior venae cavae to the right auricle. There the cycle starts again, as shown in Diagram IX–1. When there is pulmonary atresia, the course of the circulation is essentially the same as that shown in Diagram VIII–1.

PHYSIOLOGY OF THE MALFORMATION

The small size of the right ventricle and the pulmonary obstruction increase the pressure in the right ventricle, which in turn raises the pressure in the right auricle. The high pressure in the right auricle holds the foramen ovale open and a right-to-left shunt is established at the auricular level. The pulmonary stenosis protects the lungs, so that the pulmonary vascular bed is normal.

CLINICAL FINDINGS

Cyanosis is usually present at birth or shortly thereafter and increases as the ductus arteriosus undergoes obliteration.

Clubbing of the extremities occurs early.

Polycythemia develops at an early age. The red blood cell count, the amount of available hemoglobin, and the hematocrit reading are all abnormally high. If the duration of life permits, polycythemia may become extreme.

Dyspnea is common but attacks of paroxysmal dyspnea rarely occur.

Diagram IX–1

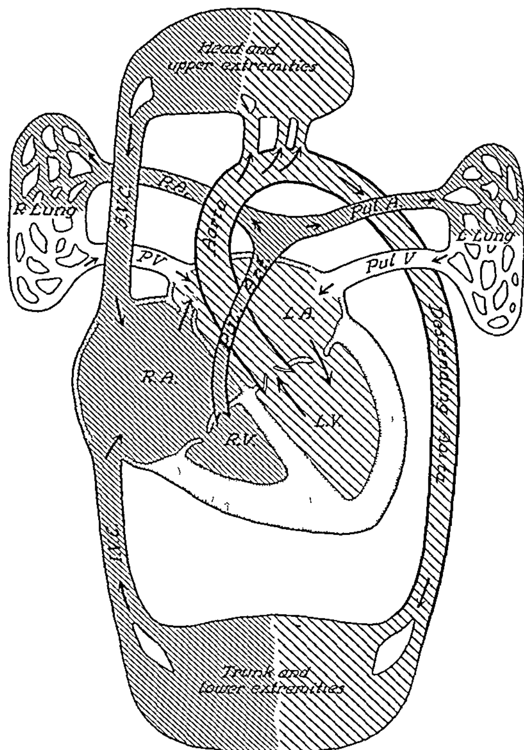
Defective development of the right ventricle with valvular pulmonary stenosis and an intact ventricular septum

The fundamental feature in this malformation is the defective development of the right ventricle, from which a small pulmonary artery arises, in addition there is usually stenosis of the pulmonary valve The ventricular septum is intact The foramen ovale is held open by the high pressure in the right auricle

Part of the blood from the right auricle flows into the small right ventricle and is pumped out through the stenosed pulmonary artery to the lungs, where it is oxygenated and returned to the left auricle in the normal manner Owing to the small size of the right ventricle, it cannot accommodate all the blood which is returned to the right auricle Hence the pressure in the right auricle rises and some blood is forced through the foramen ovale to the left auricle and mixes with the oxygenated blood returned by the pulmonary veins to the left auricle. This admixture of arterial and venous blood flows into the left ventricle and is pumped out through the aorta to the systemic circulation It is returned in the normal manner by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again

Clinical diagnosis The infant suffers from persistent cyanosis, which dates from birth but does not suffer from attacks of paroxysmal dyspnea, nevertheless the patient's incapacity becomes progressively greater as he grows older The heart is slightly to moderately enlarged with a concave curve at its base. The second sound over the pulmonary area is diminished and the pulmonary vascular markings are reduced The electrocardiogram shows only slight evidence of hypertrophy of the right ventricle in V_1 and an early transition zone to left ventricular dominance

Diagram IX–1



☐	Arterial blood (fully saturated)
▨ (broken hatching)	Small admixture of venous blood No visible cyanosis
▨	Venous and arterial blood mixed Cyanosis visible
▩	Venous blood



Left anterior-oblique position



Right anterior-oblique position

FIGURE IX-5 Defective development of the right ventricle with extreme pulmonary stenosis and an intact ventricular septum (same patient as in Figure IX-4) Infant

shadow extends abnormally far posteriorly (see Figure IX-5) The pulmonary window is clear In the right anterior-oblique position there is no enlargement of the left auricle

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram gives the clue to the diagnosis There may or may not be a right axis deviation The unipolar precordial leads always reveal less evidence of right ventricular hypertrophy than would be expected from the clinical picture There may be an R wave of moderate height in V_1, but never the evi

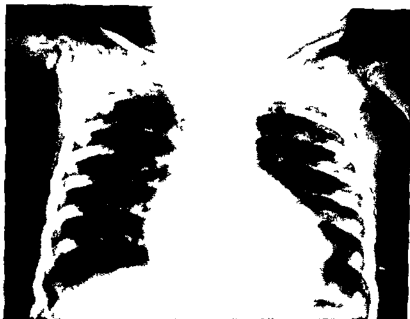


FIGURE IX-3 Defective development of the right ventricle with valvular pulmonary stenosis and an intact ventricular septum (Case IX-1) Child



FIGURE IX-4 Defective development of the right ventricle with extreme pulmonary stenosis and an intact ventricular septum (same patient as in Figures IX-5, 7) Infant



Left anterior oblique position



Right anterior-oblique position

FIGURE IX-5 Defective development of the right ventricle with extreme pulmonary stenosis and an intact ventricular septum (same patient as in Figure IX-4) Infant

shadow extends abnormally far posteriorly (see Figure IX-5) The pulmonary window is clear In the right anterior oblique position there is no enlargement of the left auricle

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram gives the clue to the diagnosis There may or may not be a right axis deviation The unipolar precordial leads always reveal less evidence of right ventricular hypertrophy than would be expected from the clinical picture There may be an R wave of moderate height in V1, but never the evi

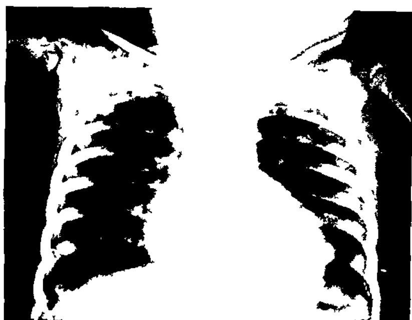


FIGURE IX-3 Defective development of the right ventricle with valvular pulmonary stenosis and an intact ventricular septum (Case IX-1) Child



FIGURE IX-4 Defective development of the right ventricle with extreme pulmonary stenosis and an intact ventricular septum (same patient as in Figures IX-5, 7) Infant

right ventricle are markedly elevated Indeed, the pressure in the right ventricle may be higher than the systemic pressure The aorta cannot be entered from the right ventricle It may not be possible to catheterize the pulmonary artery If the pulmonary artery is entered, the oxygen content will be the same as or lower than that of the right ventricle and the pressure will be low

If the patient is catheterized through the saphenous vein, it is frequently possible to catheterize the left auricle through the foramen ovale, which is held open by the high pressure in the right auricle The oxygen saturation in the left auricle the left ventricle, and the femoral artery is reduced

Angiocardiography shows a large right auricle in which the dye lingers for some time Usually the dye is seen to pass to the left auricle and the left ventricle as well as to the right ventricle The right ventricle is seen to be small and the pulmonary artery may or may not be visualized, depending on the severity of the pulmonary stenosis The aorta is not opacified until after the left auricle has filled Furthermore, in the lateral view the aorta is seen to arise in its normal position

DIAGNOSIS

The diagnosis is based upon the occurrence of intense cyanosis and increasing incapacity Although cyanosis dates from birth, a history of attacks of paroxysmal dyspnea is seldom obtained and the child rarely squats when tired When the right ventricle is tiny or when the pulmonary artery is atretic at its base, the heart enlarges rapidly and cardiac failure occurs in infancy When the condition is less severe, the malformation may simulate a tetralogy of Fallot but the electrocardiogram shows less evidence of right ventricular hypertrophy

Angiocardiography will show that the right ventricle is abnormally small and that the aorta has no connection with it Cardiac catheterization will reveal that the right ventricular pressure is abnormally high

DIFFERENTIAL DIAGNOSIS

In early infancy the condition requires differentiation from other causes of cardiac enlargement and cardiac failure, notably "pure" pulmonary stenosis of a severe degree and Ebstein s anomaly of the tricuspid valve, and also from the malformation in which both vessels arise from the right ventricle In older children the condition may be confused with a tetralogy of Fallot, truncus arteriosus with reduced pulmonary blood flow a tricuspid atresia, and possibly a single ventricle with pulmonary stenosis

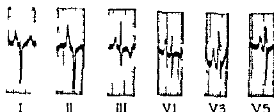


FIGURE IX-6 Defective development of the right ventricle with valvular pulmonary stenosis and an intact ventricular septum (Case IX-1) Child

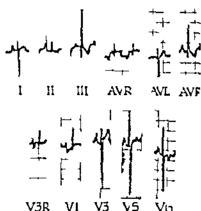


FIGURE IX-7 Defective development of the right ventricle with extreme pulmonary stenosis and an intact ventricular septum (same patient as in Figure IX-4) Infant

dence of extreme right ventricular hypertrophy such as is characteristic of severe pulmonary stenosis with an intact ventricular septum Usually the precordial leads show as much evidence of left ventricular dominance as of right ventricular hypertrophy Thus V1 may show an S wave of greater height than the R wave, in addition there is frequently an early transition zone to left ventricular dominance The P waves are tall and peaked, definitely of the Himalayan type Indeed, the combination of Himalayan P waves and relatively little evidence of right ventricular dominance in V1 (see Figures IX-6 and 7) is always suggestive of this malformation

SPECIAL TESTS

The red blood cell count, the amount of available hemoglobin and *the hematocrit reading* are all abnormally high

The oxygen saturation of the arterial blood is reduced and may be extremely low

Cardiac catheterization shows essentially the same oxygen content in the right auricle and the right ventricle The pressures in the right auricle and the

TREATMENT

When the right ventricle is abnormally small and the aorta does not over ride the ventricular septum, a systemic pulmonary anastomosis leads to right sided cardiac failure (see Illustrative Case IX-1)

A valvulotomy is of some benefit but the small size of the right ventricle prevents that chamber from carrying its full load Indeed, the right ventricle is so small that a transventricular valvulotomy is impossible, hence the operation must be done through the pulmonary artery under direct vision Unfortunately the relief of the pulmonary stenosis does not alter appreciably the size of the right ventricle, which remains abnormally small For this reason, after valvulotomy, no attempt should be made to close the auricular defect, as it acts as an escape valve for the high pressure in the auricle

The larger the right ventricle, the greater is the benefit derived from operation. Furthermore, the larger the right ventricle, the longer is the condition compatible with life For this reason the operation is usually of greater benefit to children than to infants

When the right ventricle is extremely small, Glenn's[1] operation to by pass the right ventricle by the anastomosis of the superior vena cava to the right pulmonary artery may also be of benefit

PROGNOSIS

The prognosis is poor but varies with the size of the right ventricle When a diminutive right ventricle is combined with pulmonary atresia, the prognosis is extremely poor When the right ventricle is a fair sized chamber, the condition may be compatible with life for a number of years Usually the child becomes progressively more severely incapacitated

Operation either to relieve the pulmonary stenosis or to by pass the right ventricle alleviates the condition but the abnormality in the size of the right ventricle remains a permanent handicap

SUMMARY

Defective development of the right ventricle with an intact ventricular septum places a great strain on the right side of the heart The condition is almost always associated with pulmonary stenosis, either valvular, infundibular, or both

Cyanosis dates from birth and increases in intensity Dyspnea is marked but the infant does not suffer from attacks of paroxysmal dyspnea

'Pure' pulmonary stenosis with a normal right ventricular cavity always shows far greater electrocardiographic evidence of right ventricular hypertophy in V_1 than does the malformation under discussion, by the time that cardiac failure develops the electrocardiogram usually shows the pattern of so-called right ventricular "strain"

Ebstein's anomaly of the tricuspid valve may be confused with defective development of the right ventricle and an intact ventricular septum in early infancy, because the characteristic electrocardiographic pattern of Ebstein's anomaly is frequently not present in the early months of life In Ebstein's anomaly the engorgement of the liver is not associated with pulsations at its margin and cardiac catheterization reveals a normal pressure in the right ventricle and in the pulmonary artery

The origin of both great vessels from the right ventricle causes early persistent cyanosis and may lead to progressive cardiac enlargement and cardiac failure The electrocardiogram, however, shows clear evidence of right ventricular hypertrophy in the precordial leads Angiocardiography reveals early opacification of the aorta as well as of the pulmonary artery

A tetralogy of Fallot differs from defective development of the right ventricle and an intact ventricular septum in that, when cyanosis appears early, the infant almost invariably suffers from attacks of paroxysmal dyspnea The heart is usually of normal size and the electrocardiogram shows greater evidence of right ventricular hypertrophy in V_1 Angiocardiography will show that the aorta receives blood from the right ventricle

Tricuspid atresia with pulmonary stenosis or atresia differs from the malformation under discussion in that the electrocardiogram shows a left axis deviation and evidence of left ventricular hypertrophy

A truncus arteriosus with reduced pulmonary blood flow causes the heart to assume a totally different contour The condition does not lead to progressive cardiac enlargement and cardiac failure In older children the two malformations are seldom confused

A single ventricle with pulmonary stenosis differs from the malformation under discussion in that, when pulmonary stenosis occurs in combination with a single ventricle, the infant suffers from attacks of paroxysmal dyspnea and the child squats when tired The patient, however, generally improves during early childhood, whereas when the right ventricle is abnormally small the child becomes progressively more incapacitated Furthermore, when there is but a single ventricle, angiocardiography reveals early opacification of the aorta

TREATMENT

When the right ventricle is abnormally small and the aorta does not over ride the ventricular septum, a systemic pulmonary anastomosis leads to right sided cardiac failure (see Illustrative Case IX-1)

A valvulotomy is of some benefit but the small *size of the right* ventricle prevents that chamber from carrying its full load Indeed, the right ventricle is so small that a transventricular valvulotomy is impossible, hence the operation must be done through the pulmonary artery under direct vision Unfortunately the relief of the pulmonary stenosis does *not alter appreciably* the size of the right ventricle, which remains abnormally small For this reason, after valvulotomy, no attempt should be made *to close the auricular defect, as it acts as an* escape valve for the high pressure in the auricle

The larger the right ventricle, the greater is the benefit derived from operation Furthermore, the larger the right ventricle, the longer is the condition compatible with life For this reason the operation is usually of greater benefit to children than to infants

When the right ventricle is extremely small, Glenn's[1] operation to by pass the right ventricle by the anastomosis of the superior vena cava to the right pulmonary artery may also be of benefit

PROGNOSIS

The prognosis is poor but varies with the size of the right ventricle When a diminutive right ventricle is combined with pulmonary atresia, the prognosis is extremely poor When the right ventricle is a fair sized chamber, the condition may be compatible with life for a number of years Usually the child becomes progressively more severely incapacitated

Operation either to relieve the pulmonary stenosis or to by pass the right ventricle alleviates the condition but the abnormality *in the size of the right ventricle* remains a permanent handicap

SUMMARY

Defective development of the right ventricle with an intact ventricular septum places a great strain on the right side of the heart The condition is almost always associated with pulmonary stenosis either valvular, infundibular, or both

Cyanosis dates from birth and increases in intensity Dyspnea is marked but the infant does not suffer from attacks of *paroxysmal* dyspnea

The heart undergoes progressive enlargement If the condition is severe, it leads to cardiac failure in early infancy If less severe, the heart enlarges so slowly that the cardiac findings may simulate a tetralogy of Fallot

The electrocardiogram, however, shows less evidence of right ventricular hypertrophy than is usual in a tetralogy of Fallot or in a severe pulmonary stenosis with an intact ventricular septum Nevertheless, the P waves are often of the Himalayan type

Angiocardiography shows a huge right auricle and a small right ventricle, the pulmonary artery may or may not be visualized The aorta is seen to arise in its normal posterior position

Cardiac catheterization reveals high pressure in the right auricle and in the right ventricle, if the pulmonary artery is entered, the pressure will be low

The diagnosis is based on the finding of early deep cyanosis in an infant who does not suffer from paroxysmal dyspnea, who has evidence of right sided cardiac failure, and whose electrocardiogram shows little or no evidence of right ventricular hypertrophy

The condition requires differentiation from 'pure" pulmonary stenosis, Ebstein s anomaly of the tricuspid valve, the origin of both vessels from the right ventricle, a tetralogy of Fallot with pulmonary atresia, tricuspid atresia, and occasionally from a truncus arteriosus or a single ventricle with reduced pulmonary blood flow

Operation for the relief of the pulmonary stenosis or to by pass the right ventricle lessens the incapacity of the patient but does not eliminate the basic abnormality

The prognosis is poor

Illustrative Cases

CASE IX–1 G K (Harriet Lane Home, No A–66254) White female First seen at the Cardiac Clinic in October, 1948, at six years of age with the chief complaints of cyanosis and shortness of breath

History The cyanosis had been present since birth and was increasing in intensity A murmur and a thrill were first detected at one year of age The child was short of breath on mild exertion, but she did not squat when tired and she never had attacks of paroxysmal dyspnea

Physical examination The child was small and poorly developed although intensely cyanotic, she was bright and cooperative The heart was of approximately normal size, the heart sounds were of good quality At this time no murmurs were audible and the second sound at the base was not reduplicated

Laboratory data Red blood cell count 10 million, hemoglobin 25 gm, hematocrit 85 per cent oxygen saturation of arterial blood 47 per cent

X ray The heart was slightly enlarged with a concave curve at the base to the left of the sternum and reduced hilar markings (see Figure IX-3)

Electrocardiogram There was a right axis deviation but there was *also evidence* of left as well as right ventricular hypertrophy in V_1 and the P waves were *abnormally* high (see Figure IX-6)

[illegible] ce of vomiting and the absence of attacks [illegible]

obviously suffering from reduced pulmonary blood flow [illegible] studies were deemed dangerous

Subsequent course In October 1948, a subclavian pulmonary anastomosis was performed by Dr Alfred Blalock. The child's color immediately improved but on the eighth postoperative day she developed right sided cardiac failure The heart became greatly enlarged The liver increased in size and pulsated The child developed pleural effusion, ascites, and edema Digitalis and diuretics were of no avail, repeated thoracenteses gave no relief

In January 1949 cardiac catheterization revealed the oxygen content of the blood in the superior vena cava to be 12 35 volumes per cent and that in the right auricle and right ventricle to be 13 0 and 13.27 volumes per cent respectively The pressure in the right ventricle was 110/27 mm of mercury The saturation of the arterial blood was 83 per cent. An angiocardiogram showed that the dye passed from the superior vena cava to the right auricle, where it lingered for a long time. The right ventricle was never well opacified but appeared to be small At the end of three seconds there was a suggestion of opacification of the main pulmonary artery and at the same time the aorta was faintly visualized The aorta appeared to arise from the left ventricle in its normal position

These studies indicated that the ventricular septum was intact and that the right ventricle was abnormally small In order to relieve the intractable right sided cardiac failure Dr Blalock created an auricular defect on January 29 A week later the patient developed auricular flutter with a rapid ventricular response Quinidine was tried but gave only temporary improvement The child lived for another *month and then died* of congestive cardiac failure

Autopsy No 21671 (performed by Dr Wharton) *Interest* was centered upon the heart The right auricle was tremendously dilated and *hypertrophied* Examination of the auricular septum showed that in addition *to the large operative defect two of the* margins of the foramen ovale were smoothly *covered with endocardium* thus indicating that prior to surgery the valve *covering the foramen ovale* had been patent. The tricuspid valve was *small and distorted* by the *very short* papillary muscle (see Figure IX-1) The right ventricle was very small its *wall* was approximately the same thick

ness as that of the left ventricle The cavity of the right ventricle was small but not rudimentary The ventricular septum was intact There was a valvular pulmonary stenosis The cusps were fused together, forming a conical dome with a perforation which admitted only the tip of a small probe The pulmonary artery was small but ballooned out to a circumference of 2 6 cm The anastomosis was intact and its margins smoothly healed The pulmonary veins returned normally to the left auricle, the left auricle itself was normal except for the defect in its wall The mitral valve, the left ventricle, the aortic valves, and the aorta were all normal

Final anatomical diagnosis Pulmonary stenosis with an intact ventricular septum, hypertrophy of an underdeveloped right ventricle, dilatation and hypertrophy of the right auricle and patency of the foramen ovale, a surgical auricular defect, and anastomosis of the proximal end of the right subclavian artery to the side of the right pulmonary artery

Comment Although the basic malformation resembled that usually seen in pulmonic stenosis with an intact ventricular septum, the cavity of the right ventricle was much smaller In this instance the pulmonary stenosis was secondary to the defective development of the right ventricle The right ventricle was so small that, even if the pulmonary valve had been normal, the ventricle could not have pumped a normal volume of blood to the lungs

Defective development of the right ventricle may occur in combination with a complete transposition of the great vessels and a high ventricular septal defect, as illustrated in the following case report

CASE IX–2 Baby F (Harriet Lane Home, No A–23101) White female Admitted to the Harriet Lane Home in December 1941, at two days of age because of episodes of cyanosis

History Birth was reported as normal, with only an eight hour labor The baby was cyanotic at birth and there was great difficulty in resuscitation Respirations were established in fifteen minutes but the baby became cyanotic on nursing The following day she became intensely cyanotic and was brought to the hospital

Physical examination The infant s color was fairly good when she was quiet but became intensely cyanotic when she cried The heart rate was rapid, the sounds were of good quality There were no murmurs The lungs were clear No abnormalities were noted in the abdomen The extremities showed cyanosis but no edema

Laboratory data Red blood cell count 7 1 million white blood cell count 9,960 hemoglobin 28 gm The urine showed a trace of albumin and rare red blood cells and was loaded with epithelial cells and white blood cells The Wassermann reaction was negative



FIGURE IX-8 Defective development of the right ventricle combined with complete transposition of the great vessels and pulmonary atresia (same patient as in Figure IX-9) Infant

X ray The heart was boot shaped with an absence of the fullness of the pulmonary conus and a narrow shadow at the base to the left of the sternum (see Figure IX-8)

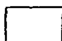
Electrocardiogram None was obtained

Clinical course The infant died at four days of age.

Clinical evaluation of the facts presented at a clinical pathological conference The contour of the heart was characteristic of a non functioning right ventricle. Neverthe less the early death of the patient pointed to a condition even more serious than that of a non functioning right ventricle with pulmonary atresia. Hence the author s diag nosis was a non functioning right ventricle combined with complete transposition of the great vessels and aortic atresia as such a malformation would cause great difficulty in the establishment of the systemic circulation

Autopsy No 1,695 (performed by Dr Sloan) The heart was small The superior vena cava and the inferior vena cava opened into the right auricle The tricuspid valve was small and opened into a diminutive right chamber The foramen ovale was cov ered by a valve but it was not sealed and had a tubular opening 0 5 cm in diameter, into the left auricle The pulmonary veins opened into the left auricle The mitral valve was normal and opened into a small but normally formed left ventricle There was a high ventricular septal defect The aorta arose from the right ventricle and par

Diagram IX–2



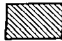
Arterial blood (fully saturated)



Venous and arterial blood
Cyanosis visible



Small admixture of venous blood
No visible cyanosis



Venous blood

Diagram IX-2

Defective development of the right ventricle with the aorta arising mainly from the right ventricle pulmonary atresia an auricular septal defect a ventricular septal defect and a patent ductus arteriosus

In this malformation the development of the right ventricle is defective the aorta is transposed and slightly over rides the ventricular septum and the pulmonary artery, which arises from the left ventricle, is atretic at its base.

The blood from the right auricle flows in part into the right ventricle and in part through the auricular septal defect into the left auricle All the blood which enters the right ventricle is pumped out into the aorta Most of the blood in the aorta is directed to the systemic circulation and returned *in the normal manner by the superior vena* cava and the inferior vena cava to the right auricle. Inasmuch as there is pulmonary atresia the pulmonary pressure remains low, some blood from the aorta will flow through the ductus arteriosus to the lungs The blood which is directed to the lungs is returned by the pulmonary veins to the left auricle. Thus the left auricle receives a

from the

o the left

ventricle The only way for the blood to be pumped out of the left ventricle is through the septal defect into the aorta which over rides the ventricular septum

In this malformation difficulty is encountered in the direction of blood to the lungs and also in the expulsion of blood from the left ventricle into the aorta

Clinical diagnosis The clinical findings are intense cyanosis and a characteristic contour of the heart The pulmonary atresia causes absence of the shadow normally cast by the pulmonary conus hence in the anterior posterior position, the shadow at the base of the heart to the left of the sternum is concave In addition in the left anterior-oblique position because of the defective development of the right ventricle, the margin of the cardiac shadow is straight

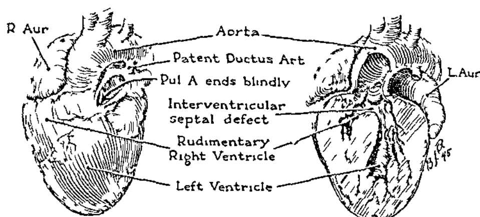


FIGURE IX-9 Defective development of the right ventricle combined with complete transposition of the great vessels and pulmonary atresia (same patient as in Figure IX-8) Infant

tially over rode the left ventricle so that blood from both ventricles was pumped out into the aorta The pulmonary artery was atretic at its base The ductus arteriosus was almost completely obliterated The course of the circulation is shown in Diagram IX-2 Figure IX-9 is a drawing of the gross specimen

Comment Although the anatomical structure was quite different from that antici pated, there was defective development of the right ventricle combined with a more serious condition than is usually found in association with a non functioning right ventricle The extreme dextroposition of the aorta rendered it difficult for the left ventricle to pump the blood out into the aorta The clarity of the lung fields should have suggested pulmonary atresia

B *Defect in the Musculature of the Right Ventricle Parchment Right Ventricle*

There is another anomaly of the right side of the heart in which the right ventricle exists as a relatively large chamber with an abnormally thin wall Two such cases have been reported in which the right ventricle was paper thin and contained virtually no heart muscle Uhl reported the case of a seven month old infant in whom autopsy showed virtually total absence of the myocardium of the right ventricle Castleman presented a similar case at one of the Massachusetts General Hospital clinical pathological conferences In Castleman s[3] case the woman lived for twenty four years and died of congestive failure with severe ascites

In this malformation, the right ventricle becomes a big, dilated, flabby cham

ber from which the blood is pumped slowly to the lungs As in all instances of abnormalities of the right side of the heart, the foramen ovale tends to be held open by the high pressure on the right side and thus a right to-left shunt is established at the auricular level

The author has seen two patients with maldevelopment of the wall of the right ventricle One was a boy of six who had shown cyanosis from birth and whose abnormality was incorrectly diagnosed as a tetralogy of Fallot Because of increasing incapacity he had had a Potts operation elsewhere He was referred to the author because of progressive cardiac enlargement and severe right sided cardiac failure The history revealed that as an infant he had never had paroxysmal dyspnea and that he did not squat when tired Autopsy revealed a large, dilated, paper thin right ventricle The myocardium of the right ventricle was virtually absent it was replaced by fat and fibrous tissue The thickened endocardium rested upon the epicardium Thus the right ventricle closely resembled the case reported by Uhl The left ventricle, however, showed some scarring and evidence of interstitial myocarditis

The other case, reported below, was carefully studied but as the patient is living, it cannot be certain that the two cases are identical *

Illustrative Case

CASE IX-3 H H (Harriet Lane Home, No B-49399) The infant was referred for diagnosis in March 1958 at the age of seventeen months

The family history was non-contributory with the only possible exception that the father was a genito urinary surgeon as was the father of one other patient with a defective development of the right ventricle †

History Growth and development were remarkably normal Except for slight persistent cyanosis the history was not significant until the day before admission, when the child had a mild convulsion and developed a left hemiplegia

Physical examination Temperature 38°C pulse 140 respirations 30, weight 8 3 kg height 73 cm The patient was well nourished and well developed There was slight persistent cyanosis which deepened on crying The left hemiplegia involved both the arm and the leg the eyes were deviated to the right The heart was enlarged The rhythm was regular There was no shock and no thrill The sounds were of fair quality and the only murmur audible was a low pitched, mid-diastolic murmur at the

* See note on page 145

† Father s profession is mentioned only because of the extraordinary coincidence that the fathers of two patients with the same unusual malformation had the same occupation

apex, such as is commonly heard in a poorly functioning heart The liver was two finger breadths below the costal margin but did not pulsate The pulses were equal in all four extremities There was no edema

Laboratory data Hemoglobin 14 3 gm hematocrit 52 per cent

Teleoroentgenogram The heart was markedly enlarged and the lung fields were clear (see Figure IX–10)

Electrocardiogram The P waves in Lead II were abnormally high and there was a left axis deviation There was no evidence of right ventricular hypertrophy, even the normal dominance of the right ventricle in V_1 was absent (see Figure IX–11)

Initial clinical diagnosis Defective development of the right ventricle

Special tests Cardiac catheterization revealed that the oxygen saturation in the superior vena cava was 33 per cent, in the inferior vena cava 23 per cent, and in the right auricle varied between 25 and 30 per cent In the three samples obtained from the right ventricle the oxygen saturation was between 30 and 40 per cent, the pressure in that chamber was 16/2 mm of mercury The pulmonary artery was not entered The left auricle and the pulmonary veins were catheterized and the blood was found to be fully saturated The pressure in the left auricle was 12/6 mm of mercury and that in the right auricle was 10/3 mm of mercury The femoral arterial saturation was 57 per cent and the pressure was 110/57 mm of mercury

The low arterial oxygen saturation showed clear evidence of a right to-left shunt It also offered the probable explanation for the occurrence of the hemiplegia, inasmuch as cerebral thrombosis may occur in infants with either polycythemia or anoxemia The fact that the left auricle was catheterized showed that the foramen ovale was patent or that there was an auricular defect Although there was a slight increase in the oxygen saturation of the blood in the right ventricle as compared with that in the right auricle, the low pressure in the right ventricle clearly excluded a significant over riding of the aorta as the source of the right to left shunt For this reason it seemed probable that the entire right to left shunt was at the auricular level and that there was also a small left to right shunt at this level

Selective angiocardiography was performed in order to exclude conclusively a ventricular septal defect First the catheter was placed in the outflow tract of the right ventricle and subsequently it was placed in the left auricle The right auricle was large, the tricuspid valve lay in its normal position The right ventricle was seen to be an enormously dilated extremely thin walled chamber (see Figure IX–12) The aorta was not visualized The great dilatation of the right ventricle beyond the tricuspid valve clearly differentiated this anomaly from Ebstein s anomaly of the tricuspid valve Furthermore, there was an area at the apex of the heart into which dye never entered The left auricle appeared normal the left ventricle was displaced upward and occupied a horizontal position (see Figure IX–13) The left ventricle, however, showed evidence



Anterior posterior position



Left anterior-oblique position

FIGURE IX-10 Defective development of the musculature of the right ventricle (Case IX-3) Infant

apex, such as is commonly heard in a poorly functioning heart The liver was two finger breadths below the costal margin but did not pulsate The pulses were equal in all four extremities There was no edema

Laboratory data Hemoglobin 14 3 gm , hematocrit 52 per cent

Teleoroentgenogram The heart was markedly enlarged and the lung fields were clear (see Figure IX–10)

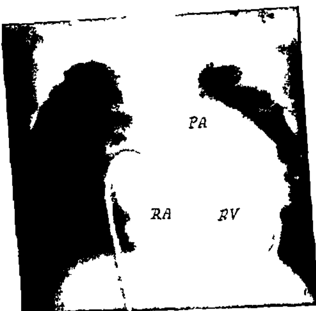
Electrocardiogram The P waves in Lead II were abnormally high and there was a left axis deviation There was no evidence of right ventricular hypertrophy, even the normal dominance of the right ventricle in V_1 was absent (see Figure IX–11)

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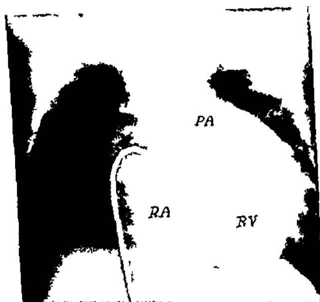


FIGURE IX-12 Defective development of the musculature of the right ventricle (Case IX-3) Infant

Dye is seen first in the pulmonary artery then in the right ventricle

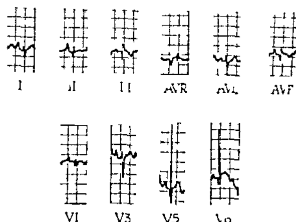


Figure IX-11 Defective development of the musculature of the right ventricle (Case IX-3) Infant

of normal ventricular contractions because there was a conspicuous difference in its size in systole and diastole

The angiocardiogram obviously did not show the exact nature of the abnormality The possibility of a tumor was considered, as it seemed well nigh impossible for a thin walled, greatly dilated chamber to displace the strong, thick walled left ventricle. A tumor in the septal wall might displace the left ventricle and either obstruct the circulation to the myocardium of the right ventricle or infiltrate its wall to such an extent as to destroy the musculature

Final clinical diagnosis Defective development of the wall of the right ventricle—a large thin walled chamber which was unable effectively to maintain the pulmonary circulation Left hemiplegia secondary to a cerebral thrombosis

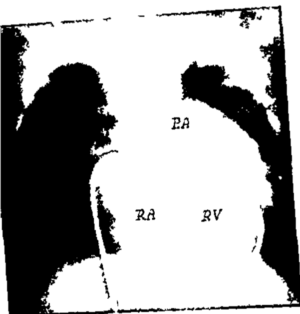
Treatment Glenn s[1] by pass operation was considered and so was vascularization of the right ventricle such as has been attempted for the left ventricle in infants in whom the left coronary artery arose anomalously from the pulmonary artery Inasmuch as either such operative procedure could be only palliative, operation was not recommended because the patient was in relatively good condition

Comment The exact nature of the malformation is not known It is however, clear that the pressure in both auricles was nearly the same as that in the right ventricle and that the right ventricle was a large chamber with such a low pressure that it was unable to pump the blood effectively to the lungs

The slight persistent cyanosis the high peaked P waves, and evidence of left ventricular preponderance in the electrocardiogram all indicated that the right ventricle, in spite of its size was seriously defective This was confirmed by the demonstration of a huge, thin walled chamber in which the pressure was low

Regardless of etiology, when the right ventricle is huge and its muscular wall is so thin that the electrocardiogram shows evidence of left ventricular dominance, the right ventricle is certainly grossly abnormal

This condition may be related to Sir William Osler s famous case of the



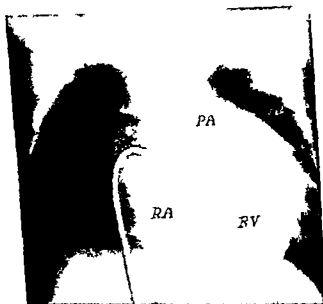
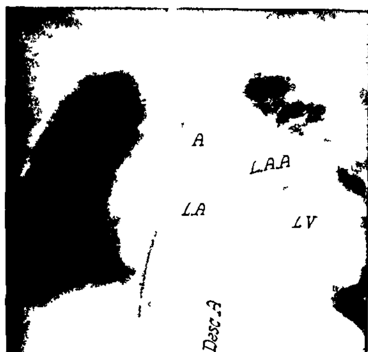


FIGURE IX-12 Defective development of the musculature of the right ventricle (Case IX-3) Infant

Dye is seen first in the pulmonary artery then in the right ventricle



Ventricular diastole



Ventricular systole

FIGURE IX–13 Defective development of the musculature of the right ventricle (Case IX–5) Infant

Dye is seen in the left side of the heart

parchment heart[4] Hence we have termed the condition a parchment right ventricle

References

1 Glenn W W L Circulatory by pass of the right side of the heart IV Shunt between superior vena cava and distal right pulmonary artery report of clinical application New England J Med 259 117-120, 1958

2 Uhl H S M Previously undescribed congenital malformation of heart almost total absence of the myocardium of right ventricle Bull Johns Hopkins Hosp 91 197-205 1952

3 Case 38201 In B Castleman and V W Towne (ed) Case Records of Massachusetts General Hospital New England J Med 246 785 1952

4 Segall H N Parchment heart (Osler) Am Heart J 40 948-950 1950

NOTE As this book was going to press the patient of Case IX-3 (see pages 139-144) had an intracardiac electrocardiogram, which showed a tracing characteristic of Ebstein s anomaly of the tricuspid valve Subsequently a Glenn by pass operation was performed but the child did not survive

Autopsy showed *extreme downward* displacement of the tricuspid valve The wall of the right ventricle was paper thin and the foramen ovale was widely patent The structure of the heart was closely similar to that shown in Figure XIX-3 (see page 470) except that no blood could enter the apical portion of the right ventricle beneath the tricuspid valve The outflow tract of the right ventricle was a thin walled readily distensible chamber Indeed the entire wall of the right ventricle was thinner than that of the right auricle Even the septal wall appeared to be involved in the abnormality for at one point it was so thin as to be translucent

Comment The annulus at the tricuspid orifice had been mistaken for the tricuspid valve in the angiocardiogram The fullness of the pulmonary conus had also been considered to be inconsistent with Ebstein s anomaly The fullness was clearly due to the dilatation of the outflow tract.

The involvement of the entire right ventricle musculature suggests that Ebstein s anomaly may be related to the parchment right ventricle except that in the latter the tricuspid valve is normal Clearly the two conditions call for differentiation In this instance the intracardiac electrocardiogram gave the clue to the diagnosis but at the time of the report we have no knowledge of the intracardiac electrocardiographic pattern of a *parchment* right ventricle

CHAPTER X

COMPLETE TRANSPOSITION OF THE GREAT VESSELS

COMPLETE transposition of the great vessels means that the aorta arises from the right ventricle and the pulmonary artery from the left ventricle This may occur as an isolated anomaly It is, however, frequently associated with an auricular or a ventricular septal defect and may be associated with almost any cardiac abnormality, as for example, a single ventricle In the latter condition the great vessels arise from a common chamber but the aorta lies anterior to the pulmonary artery

Furthermore, the pulmonary artery may be of normal size, it may be greatly dilated, or it may be stenotic or even atretic Although the same is theoretically true as regards the aorta, it is usual for the aorta to be of normal size unless the anomaly is associated with some gross malformation which dominates the clinical syndrome, such as aortic atresia

The condition is more frequently compatible with life when the pulmonary artery is greatly dilated or markedly stenosed than when both great vessels are of equal size Nevertheless, the basic principles and the underlying hemodynamics are the same in most types of transposition of the great vessels Therefore let us first consider a complete transposition of the great vessels in which both great vessels are of normal size, and then the factors which aid in the differentiation of the other types

ETIOLOGY

The etiology of all types is obscure Much evidence has been brought forward to show that the condition results from an abnormal torsion of the aortic septum whereby the aortic septum fails to meet the ventricular septum in the normal manner For a discussion of the etiology of this condition, the reader is referred to the works of Rokitansky,[1] Spitzer,[2, 3] Pernkopf and Wirtinger,[4] Harris and Farber,[5] Bremer,[6] Emerson and Green,[7] and Lev and Saphir [8]

A *Complete Transposition of the Great Vessels with Both Great Vessels of Approximately Normal Size*

NATURE OF THE MALFORMATION

The essential feature of this malformation is that the aorta arises from the right ventricle and the pulmonary artery from the left ventricle The coronary

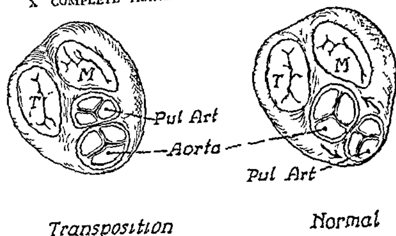


FIGURE X-1 Complete transposition of the great vessels and normal heart
The drawing shows the base of the heart with the auricles removed

arteries arise from the base of the aorta, which is usually of normal size The aorta is rotated forward and to the left and the pulmonary artery is rotated back ward and to the right in a counterclockwise direction Only rarely is the aorta rotated so far to the right that it occupies the position of the normal pulmonary artery Usually, although the aorta arises entirely from the right ventricle, it lies above the mid portion of that chamber and in most instances the pulmonary artery comes to lie directly posterior to the aorta (see Figure x-1)

Some defect within the heart, either a ventricular septal defect or an auricu lar septal defect, is generally present Indeed, in order for this malformation to be compatible with life, even for a short time, some pathway must exist which will permit at least a slight interchange of blood between the systemic and pul monary circulations If neither the ductus arteriosus nor the foramen ovale is patent, there must be an auricular or a ventricular septal defect, or an aortic septal defect, or an anomaly of the venous return, or else some combination of these anomalies

At birth the ductus arteriosus and the foramen ovale are both normally pat ent In cases of complete transposition of the great vessels in which there is no other abnormality, these pathways continue after birth to be of functional im portance and to play a vital role in the anomalous circulation Indeed, the ques- tion of prime importance to extra uterine life is whether there is the possibility of some admixture of venous and arterial blood within the auricles or within the ventricles, or whether life is dependent upon the patency of the ductus arteriosus and the foramen ovale

The structure of the heart in a complete transposition of the great vessels

with an intact ventricular septum is shown in Figure x-2 In this instance the only additional malformation which permitted the shunting of blood from one circulation to the other was a small aortic septal defect

The drawing of a complete transposition of the great vessels with a ventricular septal defect is shown in Figure x-3

COURSE OF THE CIRCULATION

During fetal life, when the great vessels are transposed, the right ventricle pumps the blood to the head and the upper extremities and some of the blood to the trunk and to the lower extremities and the left ventricle meets the high pressure of the unexpanded lungs and sends blood through the ductus arteriosus to the trunk and lower extremities Consequently the altered position of the great vessels causes no significant change in the course of the fetal circulation or in the amount of work required of the two ventricles (see Figure x-4) At birth the heart is normal in size Indeed, the condition is so inconsequential to fetal life that the baby ordinarily lives till term For this reason a complete transposition of the great vessels is a relatively common malformation

After birth the situation is quite different With the expansion of the lungs and the establishment of respiration, the left ventricle pumps the blood through the pulmonary artery to the lungs, whence it is returned in the normal fashion by the pulmonary veins to the left auricle The blood from the right ventricle is pumped out into the aorta and is returned by the superior vena cava and the inferior vena cava to the right auricle Thus there is a tendency for the blood to be pumped around and around the systemic circulation and in a similar manner around the pulmonary circulation

If there are no intracardiac abnormalities, the foramen ovale and the ductus arteriosus, which are normally patent at birth, are the only possible pathways for the crossing of the two circulations As is always the case, with the first breaths of life, blood will flow from the aorta to the pulmonary artery, the blood which is directed to the lungs is returned to the left auricle and the left ventricle Inasmuch as the valve which covers the foramen ovale closes from left to right, all the blood from the left auricle will flow into the left ventricle and from there it is again pumped to the lungs Thus the blood re circulates through the lesser circulation As long as the pulmonary pressure remains low, some blood from the aorta flows through the ductus arteriosus to the lungs Since all the blood which goes to the lungs is returned to the left side of the heart the pressure in the left ventricle and in the pulmonary artery will eventually rise

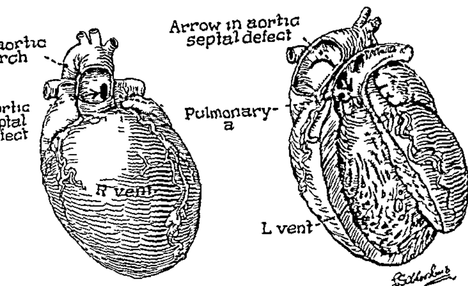


FIGURE X-2 Complete transposition of the great vessels with an intact ventricular septum and an aortic septal defect

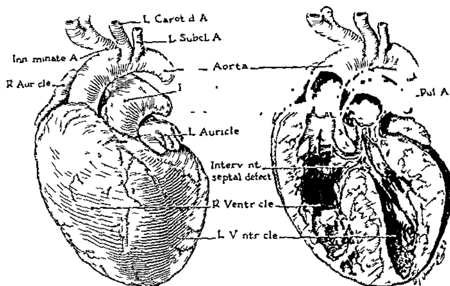


FIGURE X-3 Complete transposition of the great vessels with a high ventricular septal defect

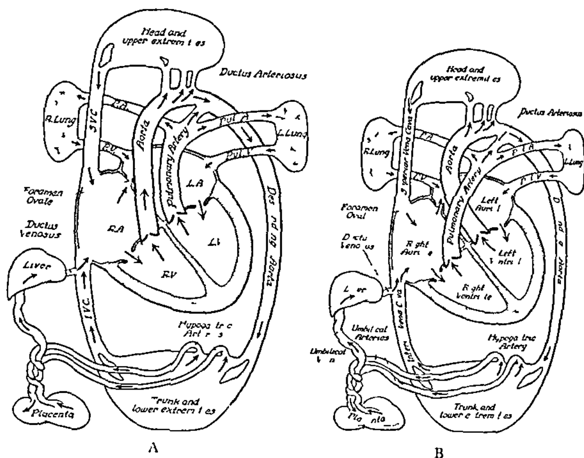


FIGURE X-4 Fetal circulation (A) Complete transposition of the great vessels with a patent ductus arteriosus and a patent foramen ovale and (B) normal heart

When the pressure in the pulmonary artery exceeds that in the aorta, the direction of the flow of blood through the ductus arteriosus will be reversed and blood will flow from the pulmonary artery to the systemic circulation The blood so shunted is returned to the right auricle Thereupon the pressure on the right side of the heart will rise, while that on the left will fall As soon as the pressure in the right auricle exceeds that in the left auricle the valve covering the foramen ovale will be forced open and blood will flow from the right auricle to the left, thereby again raising the pressure in the left auricle and the left ventricle and the pulmonary artery Thereafter blood will continue to flow from the pulmonary artery through the ductus arteriosus to the descending aorta and from the right auricle through the foramen ovale and the left auricle This mechanism, which is shown in Diagram x-1, permits the continuous crossing of the two circulations so long as the ductus arteriosus and the foramen ovale remain patent As these two pathways undergo normal obliteration, the condition becomes

incompatible with life Infants with this condition usually die within the first month

For an infant with a complete transposition of the great vessels to survive the closure of fetal pathways, there must be an additional defect, either an auricular or a ventricular septal defect or both (see Diagram x-2) or even, in rare instances, an aortic septal defect The transposition of the great vessels causes the blood which is pumped out from the right ventricle into the aorta to be returned by the superior vena cava and the inferior vena cava to the right auricle, and that which is pumped out from the left ventricle into the pulmonary artery to be returned by the pulmonary veins to the left auricle Consequently, even though a normal volume of blood reaches the lungs, difficulty is encountered in the direction of venous blood to the lungs and of oxygenated blood to the systemic circulation Hence, in this malformation, the greater the volume of the shunt, the greater is the admixture of arterial and venous blood and the less intense is the cyanosis

Furthermore, if blood is shunted from the right side of the heart to the left, it circulates through the lungs and is returned to the left side of the heart, whereas, if it is shunted from the left side to the right, it circulates through the body and is returned to the right side of the heart In other words, the blood which is shunted from one side to the other is returned to the side to which it was shunted (see Figure x-5) Consequently the blood tends to pile up on one side of the circulation There is, however, a limit to the amount of the shunt in one direction, because while the pressure on one side steadily rises, that on the other side must fall Whenever the pressure on one side exceeds that on the other the direction of the shunt will be reversed

When the blood is shunted from the lungs to the systemic circulation, it is easy to understand that the pressure in the systemic circulation will soon exceed that in the pulmonary circulation and that the direction of the shunt will be reversed Thereupon blood will be shunted from the systemic circulation to the lungs until the pressure in the pulmonary circulation exceeds that in the systemic circulation As soon as the pressure in the pulmonary circulation exceeds that in the systemic circulation blood will again be shunted to the systemic circulation When there is but a single defect, the reversal in the direction of the shunt is the only mechanism which permits the return of blood to the side *from which* it was shunted Moreover, the direction of oxygenated blood to the systemic circulation and of venous blood to the lungs is dependent upon the perpetual reversal in the direction of the shunt This mechanism of the piling up

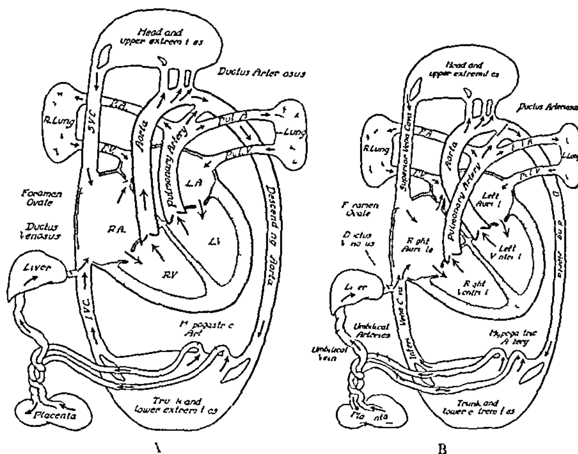


FIGURE X–4 Fetal circulation (A) Complete transposition of the great vessels with a patent ductus arteriosus and a patent foramen ovale and (B) normal heart

When the pressure in the pulmonary artery exceeds that in the aorta, the direction of the flow of blood through the ductus arteriosus will be reversed and blood will flow from the pulmonary artery to the systemic circulation. The blood so shunted is returned to the right auricle. Thereupon the pressure on the right side of the heart will rise, while that on the left will fall. As soon as the pressure in the right auricle exceeds that in the left auricle the valve covering the foramen ovale will be forced open and blood will flow from the right auricle to the left, thereby again raising the pressure in the left auricle and the left ventricle and the pulmonary artery. Thereafter blood will continue to flow from the pulmonary artery through the ductus arteriosus to the descending aorta and from the right auricle through the foramen ovale and the left auricle. This mechanism, which is shown in Diagram X–1, permits the continuous crossing of the two circulations so long as the ductus arteriosus and the foramen ovale remain patent. As these two pathways undergo normal obliteration, the condition becomes

DIAGRAM X-1

Complete transposition of the great vessels with a patent ductus arteriosus and a patent foramen ovale

the high pressure in the right auricle and permits the blood to flow in one direction only namely *from right to left*

The blood from the right auricle flows into the right ventricle, is pumped out through the aorta to the systemic circulation, and is returned by the superior and inferior venae cavae to the right auricle The blood from the left auricle flows into the left ventricle and is pumped out through the pulmonary artery to the lungs whence it is returned by the pulmonary veins to the left auricle

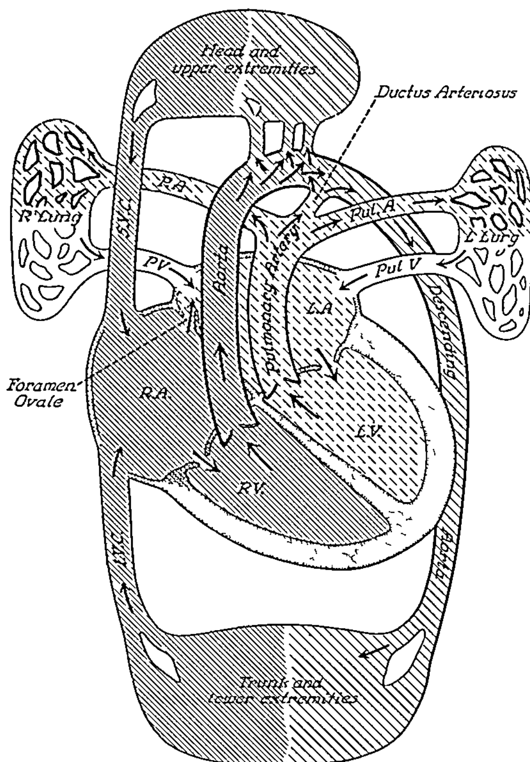
Inasmuch as the valve covering the foramen ovale closes from left to right, blood can be shunted only through the foramen ovale from right to left The blood so shunted raises the pressure in the left auricle and the left ventricle and in the pulmonary artery Therefore some blood from the pulmonary artery flows through the ductus arteriosus to the descending aorta The blood so shunted is returned to the right *auricle thus raising the pressure in the right auricle. The foramen ovale is again forced* open and some blood from the right auricle flows to the left auricle and thence to the left ventricle There the cycle starts again

Clinical diagnosis The heart becomes greatly enlarged In the *anterior posterior* position there is an absence of the shadow cast by the pulmonary conus and the *shadow* cast by the great vessels is narrow this shadow increases in width when viewed in the left anterior-oblique position Murmurs are of no diagnostic aid The electrocardiogram usually shows a right axis deviation and evidence of right ventricular hypertrophy

Cyanosis is intense Inasmuch as the direction of the flow of blood *is from the* pulmonary artery through the ductus arteriosus to the lower extremities the left ventricle *is pumping against systemic pressure consequently there is pulmonary hypertension* Furthermore some arterial blood *is directed through* the ductus arteriosus to the descending aorta hence the lower extremities *are* less cyanotic than the upper extremities The line of demarcation of the cyanosis lies at *the brim of the pelvis*

The double lines as shown in the lower right *hand block* indicate that the cyanosis is of greater intensity in the head and the upper extremities than in the lower extremities

DIAGRAM X–1



☐	Arterial blood (fully saturated)	▨	Venous and arterial blood Cyanosis visible
▨	Small admixture of venous blood No visible cyanosis	▨	Venous blood with but slight admixture of oxygenated blood
▨	Venous blood		

DIAGRAM X-2

Complete transposition of the great vessels with auricular and ventricular septal defects and a patent ductus arteriosus

In this malformation the aorta arises from the right ventricle and the pulmonary artery from the left ventricle In addition, there is a gross defect in both the auricular septum and the ventricular septum The ductus arteriosus *may remain patent*

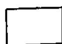
The blood from the right auricle flows into the right ventricle and can be pumped out into either the aorta or the pulmonary artery Inasmuch as the aorta arises from the right ventricle the greater part of the blood which enters the right ventricle is pumped out through the aorta to the systemic circulation All the blood *in the systemic circulation is returned* by way of the superior and inferior venae cavae to the right auricle Similarly the blood which passes from the left auricle to the left ventricle can leave by way of the pulmonary artery or by way of the aorta Inasmuch as the *pulmonary artery* arises directly from the left ventricle, most of the blood *in the left ventricle* is pumped into the pulmonary artery *This blood goes to the* lungs *for* oxygenation and the oxygenated *blood* is returned to the left auricle and thence to the left ventricle

The high ventricular septal defect *offers* easy opportunity for some oxygenated blood from the left side of the *heart to* be *pumped* out into the aorta The blood *so* shunted is returned to the right *auricle* and the right ventricle As the pressure on the right side rises blood *will* be *shunted* from *right to left* through both the auricular septal defect and the ventricular septal defect *Thereby venous* blood is directed to the lungs for oxygenation As *in all* cases of *complete* transposition of the great vessels, there is always pulmonary *hypertension In* this malformation there is ample *opportunity* for shunting blood *from* one side of the heart *to the other* and equal opportunity *for a* reversal in the direction of the shunt *The direction* of the flow of blood through the ductus arteriosus will depend upon *the relative* pressures in the aorta and the pulmonary artery However the *crossing of the two* circulations *is* not dependent *upon* this pathway Therefore the *condition is* compatible with *life* after the closure of the *ductus arteriosus* the ductus arteriosus may *or may* not be patent

Clinical diagnosis is based upon *the size and the* contour of the heart The heart is enlarged both ventricles are huge There *is absence* of fullness of the pulmonary *conus* and the shadow cast by the *great* vessels is narrow *This shadow* increases in width when *viewed in the left* anterior-oblique *position There may* be evidence *of* congestion in the lungs Murmurs are of *no* aid in *diagnosis* The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy

Diagram X–2

Head and upper extremities
Ductus Arteriosus
R Lung
L Lung
PA
Pul. A.
Pul. Art.
S.V.C.
PV
Pul V
Aorta
Pulmonary Art.
Descending
Aorta
R.A.
L.A.
R.V.
L.V.
I.V.C.
Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

is pulmonary stenosis or great dilatation of the pulmonary artery than when the pulmonary artery is of normal size If such a balance is established, although the heart is enlarged, the enlargement is not progressive and the condition may be compatible with life for a number of years (see Sections B and C)

PHYSIOLOGY OF THE MALFORMATION

In this malformation, instead of the two circulations crossing in the normal way, the blood is directed around and around the systemic circulation and similarly around the pulmonary circulation Moreover, it is venous blood which circulates through the systemic circulation and oxygenated blood which circulates through the lungs Consequently, although some crossing of the two circulations is essential for life, there is real difficulty in the direction of venous blood to the lungs The effective flow is very small Furthermore, in this malformation the greater the volume of the shunt, the greater is the volume of venous blood directed to the lungs and the greater is the volume of arterial blood which can be directed to the body Hence the greater the shunt, the less intense is the cyanosis

As previously mentioned, the reversal in the direction of the shunt occurs when the pressure in one circulation exceeds that of the other, hence a balance is established when the pressure in the pulmonary circulation approximates that in the systemic circulation In other words, severe pulmonary hypertension is the rule This finding has been repeatedly confirmed at operation Indeed, the author had thought that an end-to-end systemic pulmonary anastomosis, in which the pulmonary artery is severed, would reduce the pressure in the distal segment sufficiently so that the anastomosis might aid in the direction of blood to the lungs Dr Alfred Blalock performed several such operations It was, however, found that the back pressure in the lungs was so high that the anastomosis between the proximal end of the subclavian artery and the distal end of the pulmonary artery could not function Indeed, a systemic pulmonary anastomosis is of no avail except when there is pulmonary stenosis and the pressure in the pulmonary artery is low

Edwards[9] believes and the author agrees, that in complete transposition of the great vessels, as in malformations where both great vessels arise from the same ventricle, the resistance in the pulmonary vascular bed probably regulates the amount of blood which is directed to the two circulations Be that as it may, in complete transposition of the great vessels, except when there is also pulmonary stenosis, pulmonary hypertension is the rule

It is worthy of note that, although the pulmonary blood flow may be exces-

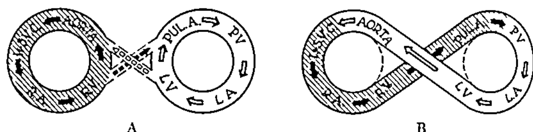


Figure X–5 (A) Course of the circulation in a complete transposition of the great vessels and (B) normal crossing of the two circulations

of blood first on one side and then on the other places an ever increasing strain on both sides of the heart, both ventricles undergo progressive dilatation and hypertrophy

The course of the circulation with a ventricular septal defect is shown in Diagram X–3 Defects in the auricular septum are usually larger than those in the ventricular septum Moreover, inasmuch as the pressure in the auricles is far lower than that in the ventricles and the difference between the pressures in the two auricles is slight, a slight increase in the pressure in either auricle represents a far greater percentage of change than a similar rise in pressure in the ventricles Therefore the crossing of the two circulations occurs more easily with an auricular septal defect than with a ventricular septal defect (see Diagram X–4) For this reason infants with a complete transposition of the great vessels usually do better when the condition is associated with a gross defect in the auricular septum than when it is associated with a ventricular septal defect

There are a few individuals with complete transposition of the great vessels in whom the defects within the heart are such that a balance is established which enables the patient to live for a number of years For this to be possible the load placed on the heart must be constant In the author's opinion such a balance is usually established by a figure of eight shunt in which the volume of the shunt from left to right is of the same magnitude as that from right to left Such a circulation can more readily be established when there are two associated anomalies than when only one is present When there is but a single defect, the crossing between the two circulations must occur through that defect and a perpetual reversal in the direction of the shunt is common When there are two defects, the shunt from left to right may be through one defect and that from right to left through the other, thereby permitting some degree of crossing of the two circulations When the two shunts are of equal volume, the load placed on the heart is constant Such a mechanism appears to be more easily established when there

Diagram X-3

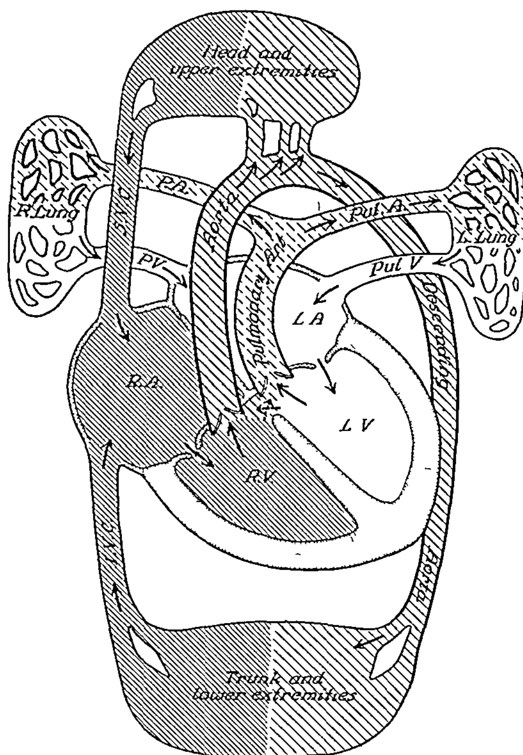
Complete transposition of the great vessels with a high ventricular septal defect

In this malformation the aorta arises from the right ventricle, the pulmonary artery from the left ventricle. There is in addition a high ventricular septal defect

The blood from the right auricle flows into the right ventricle, from there most of the blood is pumped out through the aorta to the systemic circulation and is returned by the superior and inferior venae cavae to the right auricle Part of the blood from the right ventricle is pumped through the septal defect into the pulmonary artery This venous blood from the right ventricle is mixed with the oxygenated blood from the left ventricle and flows to the lungs, where the blood is oxygenated It is, however, only the venous blood from the right ventricle which is able to take up oxygen All the blood from the lungs is returned by the pulmonary veins to the left auricle, thence it flows to the left ventricle and is again pumped out into the pulmonary artery Just as some blood from the right ventricle may pass through the septal defect to the pulmonary artery, so some blood from the left ventricle may pass through the defect into the aorta In whichever direction the blood is shunted, it is returned to that same side This mechanism tends to increase the pressure on the side to which the blood is shunted and lower the pressure on the side from which the blood is shunted Hence the relative pressure on the two sides of the heart is reversed Consequently the direction of the shunt is reversed Thus each side of the heart alternately has an excessive amount of work Both ventricles undergo progressive enlargement It is the relative pressure in the two circulations which regulates the direction and the volume of the shunt The systemic and pulmonary pressures are approximately equal, there is marked pulmonary hypertension Great difficulty however, is encountered in the direction of the oxygenated blood to the systemic circulation

Clinical diagnosis Cyanosis is intense Clubbing develops at an early age The heart undergoes progressive enlargement A systolic murmur and a gallop rhythm are the rule Cardiac failure occurs early with congestion in the lungs engorgement of the liver and edema of the extremities The contour of the heart is egg shaped with a narrow pedicle at the base which increases in width in the left anterior-oblique position The vascular markings extend nearly to the periphery of the lungs The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy

DIAGRAM X-3



Arterial blood (fully saturated)	Venous and arterial blood Cyanosis visible
Small admixture of venous blood No visible cyanosis	Venous blood

DIAGRAM X-4

Complete transposition of the great vessels with an auricular septal defect

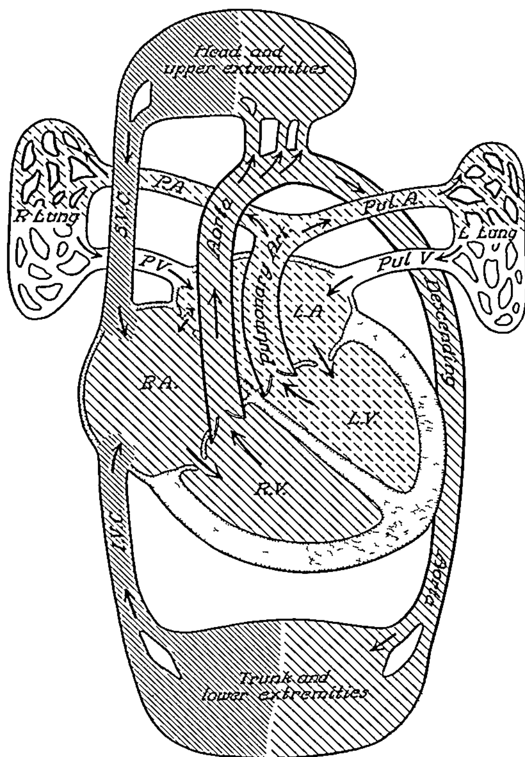
In this malformation the aorta arises from the right ventricle, the pulmonary artery from the left ventricle and there is a gross defect in the auricular septum The ductus arteriosus undergoes normal obliteration

The blood from the right auricle flows into the right ventricle and is pumped out by way of the aorta to the systemic circulation, thence it is returned by the superior and inferior venae cavae to the right auricle. The blood from the *left auricle flows into the left ventricle and is pumped out* through the pulmonary artery to the lungs whence it is returned by the pulmonary veins to the left auricle. The gross defect in the auricular septum permits the flow of blood in either direction consequently, as the lungs expand and blood flows readily from the left ventricle to the lungs and also through the ductus arteriosus to the lungs the *volume of blood which flows to the lungs is increased* This increased volume of blood is returned to the left auricle. Therefore the pressure in that chamber will rise until it exceeds that of the right auricle, whereupon the direction of the shunt will be reversed There the cycle starts again Because of the low pressure in the auricles a very slight increase or decrease in the pressure in either auricle may alter the *direction and the volume of the shunt* Such changes occur regardless of whether the ductus arteriosus is patent Nevertheless an equilibrium cannot be established until the pulmonary pressure is approximately equal to the systemic pressure Consequently there must be pulmonary hypertension

Clinical diagnosis Cyanosis is obvious but may not be as intense as it is in other types of transposition of the great vessels The heart is enlarged There is absence of fullness of the pulmonary conus In the anterior posterior position the shadow cast by the great vessels is narrow this shadow increases in width when viewed in the left anterior-oblique position Murmurs are of no diagnostic significance The electrocardiogram *shows a right axis deviation and evidence of right* ventricular hypertrophy

If the defect in the auricular septum is relatively large, the low pressure in the auricle permits the ready shunting of blood Inasmuch as any increase in the shunt in either direction is beneficial, a balance may be established with a relatively good pulmonary blood flow and proportionally less cyanosis Under such circumstances the condition may be compatible with life for a number of years

Diagram X-4



☐	Arterial blood (fully saturated)	▨	Venous and arterial blood Cyanosis visible
▧	Small admixture of venous blood No visible cyanosis	▨	Venous blood

arteries, both of which arise from the subclavian arteries, the line of demarcation of the cyanosis lies at the brim of the pelvis

This distribution of cyanosis is most conspicuous when a complete transposition of the great vessels is combined with complete interruption of the isthmus of the aorta (see Section D and Diagram X-9) It is, however, most frequently seen in young infants with a complete transposition of the great vessels in whom the intracardiac defect is small or *absent* and both the ductus arteriosus and the foramen ovale are patent

Clubbing of the extremities develops as the red blood cell count rises It is *usually apparent* by one year of age

Polycythemia steadily increases The red blood cell count may approach *10* million and there is a proportionate increase in the amount *of available hemoglobin* and in the hematocrit reading If the duration of life permits, the secondary changes in the blood usual with long standing polycythemia *develop* the blood platelets and the blood fibrinogen are decreased and the blood clot becomes friable

Respirations are rapid and shallow

Barrel shaped chest deformity develops, owing to air hunger and the excessive rapidity of the respirations

Repeated episodes of loss of consciousness may occur but attacks of paroxysmal dyspnea are rare The pressures in the two circulations are approximately equal and there is no abrupt change in the flow of blood to the lungs The blood gradually piles up first on one side and then on the other The infant suffers from progressive anoxemia until he finally loses consciousness Indeed, he may die from anoxemia unless the shunt reverses, whereupon he gradually improves

Difficulty in feeding and failure to gain are common complaints Weight gain is usually extremely slow because the infant can digest only a small amount of food at a time and simply cannot eat enough to *meet the normal requirements* of the body

Growth and development are retarded The baby is slow to hold up his head alone, to turn over, and to crawl Few of these children learn to walk before three or four years of age Their exercise tolerance is extremely limited and they squat when tired Their growth may be extremely stunted (see Section C)

Pulmonary congestion is of common occurrence unless there is an associated pulmonary stenosis Usually plenty of blood reaches the lungs Consequently, as compensation fails, the patient usually develops rales in the lungs

The liver becomes engorged, it not infrequently descends to the umbilicus

sive, over a period of years collateral circulation does develop Inasmuch as the aorta carries venous blood, such collateral circulation aids in the direction of venous blood to the lungs

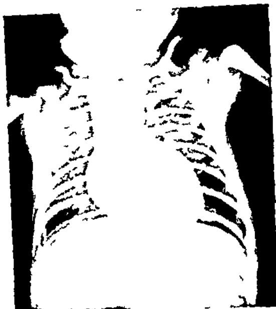
CLINICAL FINDINGS

The outstanding clinical manifestations are due to anoxemia, polycythemia, and difficulty in the direction of oxygenated blood to the systemic circulation

Cyanosis may or may not be apparent at birth Regardless of the structure of the heart, the first breaths of life direct the blood to the lungs If the cardiac septa are intact, great difficulty is encountered in the direction of the oxygenated blood to the systemic circulation Cyanosis appears early and is intense If, however, there is free communication between the two sides of the heart, that is, if there is a gross defect in either the ventricular or the auricular septum, infants with this malformation may seem remarkably normal during the neonatal period Cyanosis may be entirely absent, or it may be apparent only when the baby cries or nurses Indeed, such a baby may be discharged from the nursery as a normal infant with a normal weight gain Nevertheless, inasmuch as the right ventricle pumps the blood around and around the systemic circulation, sooner or later cyanosis almost invariably develops and becomes progressively more intense

The author has, however, seen two patients with complete transposition of the great vessels who showed no cyanosis even in childhood Both were operated on because of large ventricular septal defects In one instance there was a gross defect in the auricular septum and in the other the ventricular septum was deviated so that, although the great vessels occupied their normal positions, the pulmonary artery arose from the left ventricle and the aorta arose from the right ventricle (for the findings upon cardiac catheterization, see below under Special Tests)

The distribution of the cyanosis is often of great diagnostic significance In this malformation the blood from the left ventricle may be pumped from the pulmonary artery through the ductus arteriosus to the descending aorta Under such circumstances oxygenated blood from the left ventricle is directed to the trunk and the lower extremities (see Diagram X–1) Consequently the lower extremities will be less cyanotic than the upper The difference in cyanosis, though definite, is not conspicuous and is best appreciated by placing the infant's hand beside his foot Inasmuch as the skin of the thorax and abdomen receives its nutrition through the internal mammary arteries and the superficial epigastric



At one month



At eight and one half months

FIGURE X-6 Complete transposition of the great vessels with a high ventricular septal defect and a situs inversus Anterior posterior position shown in reverse

The reversal of the x rays makes it easier to compare the contours with those of the same malformation in a heart that occupies its normal position

and may extend to the brim of the pelvis There is seldom either tricuspid in sufficiency or stenosis, hence the liver does not pulsate

Edema of the extremities occurs with cardiac failure

The pulse pressure is narrow, *the blood pressure* is often difficult to obtain

CARDIAC FINDINGS

The heart at birth is normal in size Unless the duration of life is too short to permit the heart to enlarge, the malformation usually leads to progressive car diac enlargement The greater the difficulty in the crossing of the two circula tions, the sooner the signs of distress become apparent and the more rapidly the heart enlarges Inasmuch as the right ventricle pumps the blood throughout the systemic circulation, that ventricle becomes huge The left ventricle also carries a full load It pumps the blood around and around the pulmonary circulation under abnormally high pressure, in addition, while the ductus arteriosus re mains patent, the left ventricle also pumps some blood through the ductus ar teriosus to the descending aorta Hence the left ventricle also becomes greatly enlarged The rate at which the heart enlarges varies with the associated anoma lies In most instances, however, the heart becomes enormously enlarged be tween seven and ten months of age, as illustrated in Figures X-6 and 7 Figures X-8 and 9 show the oblique views of the same infant

Nevertheless, occasionally a balance is established, cardiac enlargement comes to an end Although theoretically this may occur after any degree of enlargement, the patient is far more likely to survive if a balance is established promptly Consequently in patients who live to childhood and early adult life the heart is usually only slightly enlarged

The heart sounds are forceful The *second sound* over the pulmonary area is usually accentuated, as the aorta lies far to the left

Murmurs and thrills are of no diagnostic importance The quality and inten sity of both the murmur and the thrill vary with the nature of the concomitant malformation and also with the relative pressure in the two circulations Usually the systolic murmur is not very intense Inasmuch as the pressures in the two circulations are approximately equal, the systolic murmur generally lacks the rasping quality characteristic of a ventricular septal defect For the same reason, even if the ductus arteriosus is patent, a continuous murmur never develops

A thrill may or may not be palpable over the precordium

A gallop rhythm is frequently audible, as compensation is usually precarious

Cardiac failure is common In most instances, the heart undergoes progres



At one month



At eight and one half months

FIGURE X-6 Complete transposition of the great vessels with a high ventricular septal defect and a situs inversus Anterior posterior position shown in reverse

The reversal of the x rays makes it easier to compare the contours with those of the same malformation in a heart that occupies its normal position

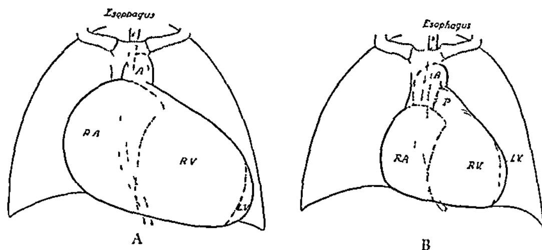


FIGURE X-7 (A) Complete transposition of the great vessels with a high ventricular septal defect and (B) normal heart Infant

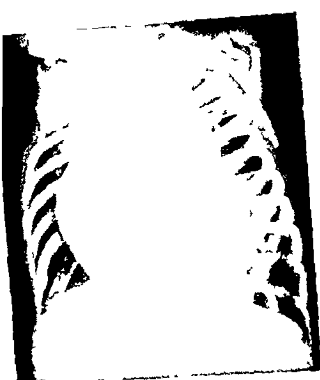
sive enlargement and eventually becomes enormous The liver is enlarged and, inasmuch as the circulation to the lungs is adequate, pulmonary congestion and rales in the lungs are common Indeed, this is the outstanding malformation of early infancy in which one finds persistent cyanosis and cardiac failure with en gorgement of the liver, edema of the extremities, and congestion of the lungs These infants usually die before eighteen months of age Death is due either to anoxia or cardiac failure combined with pulmonary congestion

X RAY AND FLUOROSCOPIC FINDINGS

The contour of the heart varies with the age of the patient and the size of the pulmonary artery Usually all four chambers of the heart are greatly enlarged The enlargement extends both to the right and to the left

In infancy the contour of the heart frequently resembles an egg lying on its side with the tip pointing slightly downward and to the left, the base of the egg, which lies to the right of the sternum, is formed by the dilatation of the right auricle (see Figure X-6, also Figures X-10 and 11) Although the aorta arises from the right ventricle, it seldom arises as far to the left as does the normal pul monary artery Hence in the anterior posterior position it is usual to find a slight concavity rather than a convexity of the upper border of the cardiac silhouette to the left of the sternum

In the left anterior oblique position both ventricles are seen to be enlarged In an estimation of the size of the right ventricle, it is important to remember that in this malformation the aorta is displaced anteriorly For this reason the in crease in the size of the right ventricle relative to the aorta is less pronounced



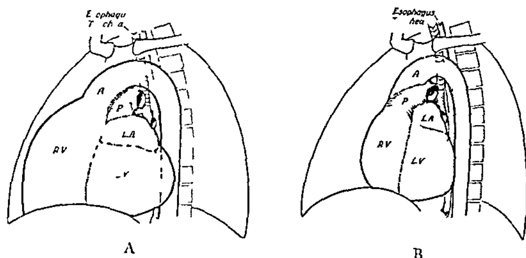
Right anterior-oblique position shown in reverse



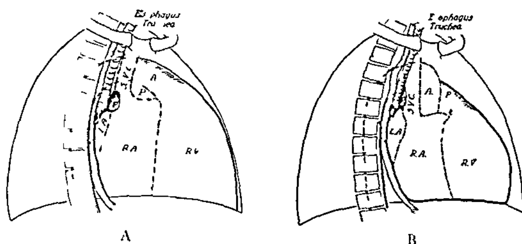
Left anterior-oblique position shown in reverse

FIGURE X-8 Complete *transposition* of the great vessels with a high ventricular septal defect and a situs inversus (same patient as Figure x-6) At eight and one half months

The reversal of the x rays makes it easier to compare the contours with those of the same malformation in a heart that occupies its normal position



LEFT ANTERIOR-OBLIQUE POSITION



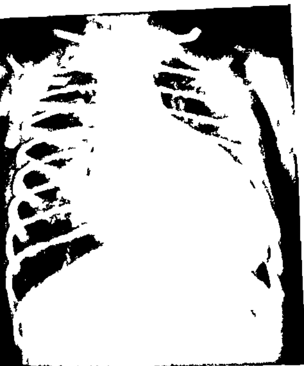
RIGHT ANTERIOR OBLIQUE POSITION

FIGURE X-9. (A) Complete transposition of the great vessels with a high ventricular septal defect and (B) normal heart. Infant.

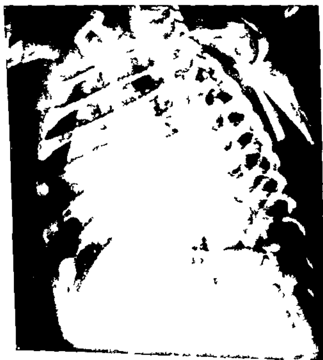
than in other malformations. Both the aorta and the right ventricle, however, extend closer to the anterior chest wall than in the normal heart (see Figures X-8 through 11). Terminally it is common to find that the right ventricle nearly touches the anterior chest wall and that, when the patient is rotated to an angle of 60°, the left ventricle extends posterior to the spinal column.

In the right anterior oblique position the heart frequently appears to fill the entire chest. The entire esophagus may be displaced backward but there is no specific enlargement of the left auricle (see Figures X-8 and 9).

The altered positions of the great vessels relative to the ventricles cause a characteristic change in the shadow cast by these vessels.[10] When the aorta arises



Anterior posterior position



Left anterior-oblique position

FIGURE X-10 Complete transposition of the great vessels with a patent ductus arteriosus and a patent foramen ovale Infant





Anterior posterior position



Left anterior oblique position

FIGURE X-11 Complete transposition of the great vessels with a high ventricular septal defect Infant

[illegible] pulmonary artery from the left ventricle, the [illegible]

pulmonary artery lies posteriorly and further [illegible] this *counterclockwise* rotation the pulmonary artery comes to lies behind the aorta, consequently, unless the pulmonary artery is abnormally large, the shadow cast by the great vessels is narrow Upon rotation of the infant into the left anterior-oblique position, the pulmonary artery, which in the anterior posterior position lies behind the aorta, comes to lie parallel to the aorta and thereby causes the *shadow cast* by these vessels to increase in width By placing one finger behind the other and then rotating the hand 45°, the change in the width of the shadow cast by the great vessels can be clearly demonstrated Owing to the *patient s* lack of cooperation, satisfactory *roentgenograms in* infancy, especially *in the oblique positions*, are difficult to obtain For this reason fluoroscopic examination is superior

Dilatation of the superior vena cava which is of common occurrence in cardiac failure, may cause the shadow at the base of the heart to appear abnormally wide The shadow cast by the superior vena cava always lies to the right of the sternum It can be differentiated from that of the great vessels by the rotation of the infant s head to the left This maneuver causes the shadow cast by the great vessels to shift to the left but never appreciably alters the shadow of the superior vena cava

Rhythmic changes in the size of the right auricle should be sought for, because when present they are of diagnostic significance This is one of the few malformations which permit such variations, owing to the piling up of blood on one side of the heart As the blood piles up in the right auricle, that chamber will gradually distend until the pressure in the right auricle becomes sufficiently greater than that in the left auricle to force open the valve of the foramen ovale Thereupon the blood will flow from the right auricle to the left, the right auricle will collapse, and the high pressure in the left auricle will close the valve covering the foramen ovale Thereafter the pressure in the right auricle will steadily rise until its pressure again exceeds that in the left auricle Then the valve will open again and the process will be repeated *This causes a rhythmic change in* the size of the right auricle which is independent of the heart rate This type of change is indicative of a malformation which causes the blood to accumulate in the right auricle, combined with a foramen ovale which is covered by a valve that is not completely sealed and permits the flow of blood in only one direction, namely, from right to left

Alteration in the contour of the heart associated with the growth of the patient If the nature of the concomitant malformation is such that the condition does not cause progressive cardiac enlargement, a circulatory balance may be established, and the patient may live for a number of years As previously stated, this is more likely to occur if there is pulmonary stenosis or great dilatation of the pulmonary artery than if both great vessels are of normal size If such a balance is possible, it usually occurs before the heart is enormously enlarged As the child grows and the diaphragm descends, the heart occupies a more vertical position In the anterior posterior position the contour of the heart has a concave curve at the base, owing to the posterior location of the pulmonary artery (see Figure x-12) Furthermore, in the left anterior-oblique position the shadow cast by the great vessels no longer increases in width As the heart drops down, the pulmonary artery, which lies posterior to the aorta, extends to the lungs more horizontally than it does in the normal heart The consequence is that the pulmonary artery lies at an abnormally low level and the pulmonary window becomes abnormally clear For this reason the contour of the heart in the anterior posterior position and in the oblique positions may closely resemble that of a tetralogy of Fallot



FIGURE X-12 Complete transposition of the great vessels with pulmonary stenosis Child

The vascular markings are of great diagnostic importance Most malformations which cause persistent cyanosis in early infancy are associated with a great reduction in the pulmonary blood flow and excessively clear lung fields In a complete transposition of the great vessels, the volume of the pulmonary blood flow is normal, the vascular markings in infancy are normal or slightly increased and extend from the hilar region nearly to the periphery of the lungs

In childhood the vascular markings are distinctive in the x ray film Even when there is pulmonary stenosis, the pulmonary artery is a fair sized vessel There are many small, discrete, circular shadows due to blood vessels which are viewed on end as they course from the posteriorly placed pulmonary artery to the anterior portion of the lungs Moreover, the vascular markings extend far out into the lung fields, indeed nearly to the periphery of the lungs Although the pulmonary arteries are seen to pulsate vigorously at operation, the pulsations are rarely visible upon fluoroscopy This is due in part to the fact that the pulmonary arteries lie abnormally deep within the chest, so that there is a considerable amount of lung tissue superimposed upon them, and in part because, even though the pressure within the pulmonary arteries is high, the pulse pressure in these arteries is not great Usually fluoroscopy does not reveal the details of these shadows and only the dense and numerous shadows in the hilar regions are visible Consequently the shadows cast by the small branches of pulmonary arteries viewed on end may be mistaken for those due to extensive collateral circulation by way of the posterior mediastinal vessels Not infrequently it is the discrepancy between the vascular shadows seen on the x ray film and those discerned upon fluoroscopy which offers the clue to correct diagnosis

In an older patient, upon slow rotation during fluoroscopic examination, it is usually possible to visualize the main branches of the pulmonary artery as they course to the lungs This may be possible in either oblique position but the left pulmonary artery is generally more readily seen in the left anterior-oblique position than in the right Indeed during fluoroscopy the lower branches of the left pulmonary artery are often visible as the patient is slowly rotated toward the left anterior-oblique position

Pulmonary stenosis renders the vascular markings even more closely *similar* to those of a tetralogy of Fallot Nevertheless, since the pulmonary artery arises from the left ventricle, the pulmonary stenosis is almost invariably valvular and the pulmonary artery is a fair sized vessel Consequently the vascular markings are heavier and extend further to the periphery of the lungs than in a tetralogy of Fallot So few patients who have a complete transposition of the great vessels

as a large ventricular septal defect because of failure to appreciate the significance of a femoral arterial saturation of 94 per cent Although in an Eisenmenger complex the saturation in the aorta may be the same as it is in the pulmonary artery, when the oxygen saturation is lower in the aorta than in the pulmonary artery, the possibility of a complete transposition of the great vessels should always be considered

Angiocardiography may be of aid in that it will demonstrate early opacification of the aorta and may or may not demonstrate late but extensive opacification of the pulmonary vascular bed (see Figure x–14) Unless the aorta is grossly misplaced (see Section c), angiocardiography is not of great help in the differentiation of dextroposition of the aorta from transposition of the aorta, and therefore *the test is seldom of aid in the differentiation of transposition of the great vessels from a tetralogy of Fallot* Occasionally in the lateral view, when both the aorta and the pulmonary artery are visualized simultaneously, the pulmonary artery can be seen to arise posterior to the aorta, thus proving that the great vessels are transposed (see Figure x–22)

DIAGNOSIS

The diagnosis is based upon the finding of a small, scrawny infant with cyanosis, rapid respirations, and a barrel shaped chest, who suffers from increasing cardiac difficulty associated with progressive cardiac enlargement, a gallop rhythm, engorgement of the liver, and congestion in the lungs When, in addition, the hands are more cyanotic than the feet, the diagnosis can be made with assurance

The x ray and fluoroscopic findings of an egg shaped heart with a narrow shadow at its base, which increases in width in the oblique view, is strong confirmatory evidence of a complete transposition of the great vessels

Children and young adults show deep cyanosis, marked clubbing of the fingers and the toes, and extreme stunting of growth The contour of the heart is similar to that of a tetralogy of Fallot The second sound over the pulmonary area is accentuated X ray findings of cardiac enlargement and a concave curve at the base of the heart to the left of the sternum, combined with dense vascular shadows which extend to the periphery of the lungs, are diagnostic of this condition

DIFFERENTIAL DIAGNOSIS

This malformation must be differentiated from a pure pulmonary stenosis, from a tetralogy of Fallot, from a single ventricle with complete transposition of

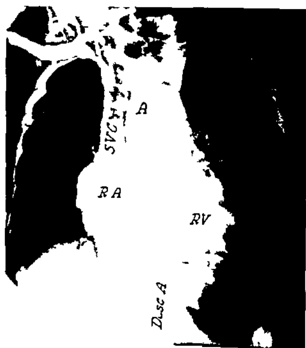




Figure X-14 Complete transposition of the great vessels Infant

the great vessels, and also from a truncus arteriosus with reduced pulm blood flow and a defective development of the right ventricle with or wi tricuspid atresia

In the neonatal period the malformation may be confused with a *'pure' monary stenosis* Confusion arises because in a patient with a "pure" pulm stenosis the foramen ovale is often functionally patent during the first few v of life Therefore the infant may appear cyanotic Poststenotic dilatation ha yet developed Consequently there is no fullness of the pulmonary conus I dition, in both malformations the vascular markings are more conspicuous in other types of pulmonary stenosis Furthermore, occasionally the infant suffer from right sided cardiac failure in the neonatal period The second s at the base of the heart to the left of the sternum is of aid in the differenti of the two malformations It is weak or absent in a baby with 'pure pulmo stenosis and accentuated when there is a complete transposition of the great sels Time is also a great aid in the differentiation of these two malformat An infant with a "pure" pulmonary stenosis usually responds to digitalis cyanosis disappears, and he does better than might be expected, whereas in malformation under discussion, if the infant is in severe difficulty during first days of life, he seldom, if ever, survives more than a few weeks

In early infancy *a tetralogy of Fallot with anatomical or functional pul monary atresia* may be confused with a complete transposition of the great sels combined with an intact ventricular septum and minimal communica between the two sides At birth, in both instances, cyanosis may be intense the heart is normal in size The outstanding difference between the two mal mations is that in a complete transposition of the great vessels the lung fi show normal or increased vascularity, whereas in a tetralogy of Fallot with p monary atresia the lung fields are phenomenally clear Further, in a comp transposition of the great vessels the heart usually enlarges rapidly Indeed, wl the condition is such as to cause difficulty in the neonatal period, the enlar ment can usually be detected within four to six days after birth, whereas in tetralogy of Fallot, the heart remains small

In older children *a tetralogy of Fallot* with a severe degree of pulmonary s nosis may be confused with a complete transposition of the great vessels in whi a balance is established and the heart is not greatly enlarged A tetralogy of F lot causes less stunting of growth, there is usually left sided chest deformity an the heart is often abnormally small, the hilar shadows may appear dense but t shadows do not extend far out into the lungs In contrast to this, a comple

transposition of the great vessels causes extreme stunting of growth, the chest is usually barrel shaped, the heart is slightly enlarged, and the vascular markings extend far out to the periphery of the lungs (see Section c)

A single ventricle with transposition of the great vessels may be confused with a complete transposition of the great vessels and a normal ventricular septum, especially when the pulmonary artery is abnormally small Clinically a patient with a single ventricle does better than one with a complete transposition of the great vessels and two ventricles, as the single ventricle permits a good opportunity for some crossing between the venous and the arterial circulation Nevertheless, it may require cardiac catheterization or angiocardiography to determine whether or not there are two ventricles

A truncus arteriosus causes great enlargement of both ventricles Although the enlargement occurs early, it is not progressive The aortic knob is usually conspicuous If the infant lives to childhood, a continuous murmur is usually audible over some portion of the lung

Defective development of the right ventricle with tricuspid atresia is characterized by clear lung fields and electrocardiographic evidence of a left axis deviation and left ventricular hypertrophy

Defective development of the right ventricle with pulmonary stenosis and an intact ventricular septum causes cardiac enlargement and great reduction in the pulmonary blood flow The second sound at the base of the heart to the left of the sternum is diminished and upon x ray the vascular markings are decreased Cardiac catheterization will show a high pressure in the right ventricle but the aorta cannot be entered Angiocardiography shows that the pulmonary artery, not the aorta arises from the right ventricle

TREATMENT

Digitalis may prolong the life of the infant

The inhalation of oxygen may be of temporary benefit

Surgical treatment has not yet been perfected The creation of an auricular septal defect, as developed by Blalock [12] has been of benefit to some patients The best results have been seen in children over two years of age These patients have shown an increase in their exercise tolerance and over a period of years some reduction has occurred in the red blood cell count, the amount of available hemoglobin and the hematocrit level Thus although the immediate results are seldom dramatic the long time results have been better than anticipated Nevertheless, this operation does not relieve the pulmonary hypertension

Baffes[13] has developed an operation for the partial transposition of the pulmonary veins to compensate for the transposition of the great vessels In this operation the right pulmonary veins are transplanted into the right auricle and a graft is inserted between the inferior vena cava and the left auricle The improvement after this operation is dramatic Initially the best results were obtained in children over two years of age, recently excellent results have also been obtained in young infants

Dr Henry Bahnson has performed one operation in which he altered the position of the auricular septum, so that the pulmonary veins drained into the right auricle and the inferior vena cava into the left auricle This operation may prove to be of great benefit

PROGNOSIS

The prognosis is poor Most infants, when the auricular and ventricular septa are closed, die of anoxemia during the first weeks of life, some infants live for a few months but the condition usually leads to progressive cardiac enlargement These infants usually die of anoxemia or cardiac failure between six and fifteen months of age Only occasionally is a balance established which permits the patient to live until childhood or early adult life

Operation definitely improves the prognosis As of 1960 the greatest improvement has resulted from the Baffes operation

SUMMARY

A complete transposition of the great vessels means that the aorta arises from the right ventricle and the pulmonary artery from the left ventricle It is a relatively common malformation, as it places no strain on the fetal circulation

After birth the blood is pumped around and around the systemic circulation and around and around the pulmonary circulation Great difficulty is encountered in the crossing of the two circulations The larger the shunt, the less intense is the cyanosis Nevertheless, the blood which is shunted from one side of the heart to the other tends to pile up on the side to which it is shunted In order to reverse the shunt, the relative pressure in the two circulations must be reversed This means that the pulmonary pressure must be approximately the same as the systemic pressure Pulmonary hypertension is the rule

Cyanosis may not be apparent during the first weeks of life, sooner or later it always develops and eventually becomes intense The distribution of the cyanosis may give a clue to the diagnosis

Clubbing develops early

Polycyth [illegible] become extreme

An infa[illegible]

may suffer from episodes of loss of consciousness [illegible]

dyspnea are ra[illegible] and shallow The chest is generally

barrel shaped [illegible]

Stunting of growth may be extreme The exercise tolera[illegible]

The child may squat when tired

The heart usually undergoes progressive enlargement As it enlarges, it develops a characteristic contour Both ventricles are always markedly enlarged There is absence of fullness of the pulmonary conus In infancy the shadow cast by the great vessels is narrow in the anterior posterior position and upon rotation of the patient into the left anterior-oblique position the shadow cast by the great vessels increases in width Inasmuch as there is adequate circulation to the lungs, the infant is likely to suffer from pulmonary congestion Death results from anoxemia or from cardiac failure with congestion in the lungs and edema of the extremities

Occasionally the malformation is compatible with life for a number of years Under such circumstances the contour of the heart and both the physical and the physiological findings closely resemble a tetralogy of Fallot The two features which differentiate this malformation from that of a tetralogy of Fallot are the accentuation of the second sound at the base of the heart to the left of the sternum and the density of the vascular markings which extend from the hilar region to the periphery of the lungs

The electrocardiogram ordinarily shows a right axis deviation and evidence of hypertrophy of both ventricles

The circulation time (arm to tongue) is abnormally short

The oxygen saturation of the arterial blood is abnormally low

Cardiac catheterization is of no great aid in the diagnosis of a complete transposition of the great vessels If the pulmonary artery is entered, the oxygen saturation of the blood in the pulmonary artery is always found to be higher than that in the femoral artery thus catheterization may occasionally give a clue to the existence of a transposition of the great vessels Cardiac catheterization may also aid in the diagnosis of the nature of the concomitant malformation

Angiocardiography shows early dense opacification of the aorta In the lateral series of films it may be possible to see that the aorta lies anterior to the pulmonary artery

The diagnosis of a complete transposition of the great vessels is based upon the finding of persistent cyanosis, increasing cardiac enlargement, and cardiac failure with engorgement of the liver and congestion of the lungs When, in addition, the hands are more cyanotic than the feet, the diagnosis can be made with assurance The diagnosis is further confirmed by the x ray evidence of a concavity at the base of the heart, absence of the shadow cast by the pulmonary conus, a narrow pedicle in the anterior-posterior position, which increases in width in the left anterior-oblique position, and dense hilar shadows with extensive vascular markings

The malformation requires differentiation from a 'pure" pulmonary stenosis, from a tetralogy of Fallot, from a single ventricle, and occasionally from a truncus arteriosus with reduced pulmonary blood flow

The creation of an auricular defect may aid in the crossing of the two circulations but does not lessen the pulmonary hypertension The Baffes operation offers greater hope for improvement of the circulation

The prognosis is poor Most infants with this malformation die between six and fifteen months of age Without operation only rarely does a patient live to adolescence or early adult life

B *Complete Transposition of the Great Vessels Combined with Enormous Dilatation of the Pulmonary Artery*

NATURE OF THE MALFORMATION

In this malformation the aorta, which arises from the right ventricle, is usually of normal size and the pulmonary artery, which arises from the left ventricle, is huge When the pulmonary artery is tremendously enlarged, it frequently over rides the ventricular septum, under such circumstances a high ventricular septal defect is an integral part of the malformation (see Figures X-15 and 16) It is, however, important to appreciate that a complete transposition of the great vessels combined with enormous dilatation of the pulmonary artery may occur in combination with only a small ventricular septal defect or even an intact ventricular septum

COURSE OF THE CIRCULATION

The course of the circulation differs from that in other cases of transposition of the great vessels only if the pulmonary artery arises in part from the right

Pulmonary a.
RA
L auricle
Ventricular septal defect
Tricuspid v
R. ventricle
L ventricle
Mitral v

FIGURE X-15 Complete transposition of the great vessels with an enormously dilated pulmonary artery

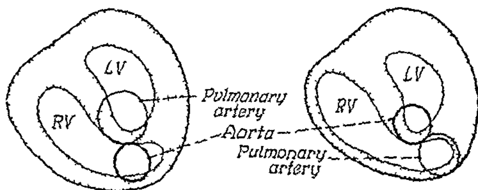


FIGURE X-16 Complete transposition of the great vessels with a dilated pulmonary artery and normal heart

ventricle Under such circumstances the blood in the right ventricle is pumped out through the aorta to the systemic circulation and through the pulmonary artery to the lungs The blood which is directed to the systemic circulation is returned by the superior and inferior venae cavae to the right auricle and that which goes to the lungs is returned by the pulmonary veins to the left auricle and thence to the left ventricle Most of the blood from the left ventricle is pumped out into the pulmonary artery When the pulmonary artery over rides the ventricular septum, some venous blood from the right ventricle is readily shunted to the lungs, and consequently an increased volume of blood is returned to the left side of the heart, which aids in the shunting of oxygenated blood across to the right side of the heart Thus some crossing of the circulation can occur, as shown in Diagram x–5 For this reason, it is not surprising to find that this combination of anomalies is frequently more compatible with life than when both great vessels are of normal size and there is no over riding of the pulmonary artery

PHYSIOLOGY OF THE MALFORMATION

The physiology of this malformation is essentially the same as that when both great vessels are of normal size, except that the volume of the pulmonary blood flow is greater When the pulmonary artery over rides the ventricular septum, the condition is physiologically and functionally similar to that of a Taussig Bing malformation, as the pulmonary artery is biventricular in origin (see Chapter xi) Nevertheless, when the pulmonary artery lies posterior to the aorta, it receives mainly oxygenated blood from the left ventricle, whereas when it lies anterior and to the left of the aorta it receives more venous blood As in all cases of complete transposition of the great vessels, it is the relative pressure in the two circulations which regulates the volume of the shunt Only when pulmonary pressure is greater than systemic pressure is oxygenated blood directed to the aorta It follows that, regardless of whether or not the pulmonary artery over rides the ventricular septum, there is always pulmonary hypertension

CLINICAL FINDINGS

Cyanosis is usually present at birth Inasmuch as the aorta arises from the right ventricle, cyanosis almost invariably appears early and eventually becomes intense Owing to the fact that the shunt is within the heart, the cyanosis is of uniform distribution

Clubbing of the extremities appears at an early age and may become extreme

Polycythemia increases with age and eventually becomes extreme As the patient grows older the usual changes in the clotting mechanism occur In late childhood or early adolescence, petechiae and blotchy purpuric eruption frequently occur

Respirations are rapid and shallow, and the chest becomes barrel shaped These infants may suffer from prolonged periods of unconsciousness due to extreme anoxemia

Difficulty in feeding, failure to gain and stunting of growth occur, as in all cases of complete transposition of the great vessels

Exercise tolerance is limited Children may squat when tired

Pulmonary congestion is common, owing to the excessive pulmonary blood flow Furthermore, the excessive pulmonary circulation renders the individual very susceptible to bronchitis and pneumonia Indeed, it is frequently difficult to determine whether the pulmonary congestion is due to cardiac failure or to infection

Hepatomegaly is the rule Even though a balance is established, there is usually slight to moderate engorgement of the liver

CARDIAC FINDINGS

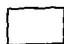
The heart is always slightly enlarged A balance is established early in life and

A systolic murmur is the rule Ordinarily the *murmur* is not very harsh Frequently a *blurred third heart sound* or a *mid diastolic murmur* is audible just inside the apex

X RAY AND FLUOROSCOPIC FINDINGS

The heart is usually slightly enlarged and there is a concave curve at its base to the left of the sternum In early infancy the vascular markings are increased but may not be conspicuous (see Figure x–17) Nevertheless, owing to the great dilatation of the pulmonary artery, as the patient grows, the hilar markings become exaggerated (see Figure x–18) Usually there are large, blotchy hilar shadows which may or may not pulsate The pulsations in these vessels are often less conspicuous than would be expected from their size, because, as previously mentioned, although the pulmonary blood flow is excessive, both the systolic and the diastolic pressure in the pulmonary artery are high, and consequently the pulse pressure in the pulmonary artery is not wide In older children, however, a hilar

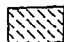
Diagram X-5



Arterial blood (fully saturated)



Venous and arterial blood
Cyanosis visible



Small admixture of venous blood
No visible cyanosis

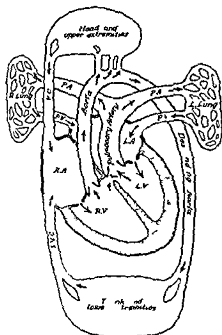


Venous blood

Diagram X-5

Complete transposition of the great vessels with a greatly dilated pulmonary artery

In this malformation a complete transposition of the great vessels is combined with a greatly dilated pulmonary artery, which frequently over rides the ventricular septum and thus receives blood from both ventricles In some instances, however, the pulmonary artery does not over ride the ventricular septum (see insert)



The blood from the right auricle flows into the right ventricle. Inasmuch as the aorta arises from the right ventricle, most of the blood in the right ventricle is pumped directly out through the aorta to the body and returned by the superior and inferior venae cavae to the right auricle, thence it again flows into the right ventricle. If the large pulmonary artery arises in part from the right ventricle some blood is pumped into the pulmonary artery, or if the pres sure is lower in the lungs, blood is shunted from the right side to the left All of the blood so shunted and most of the blood from the left ventricle is pumped out through the pulmonary artery to the lungs, where it is fully oxygenated and returned by the pulmonary veins to the left auricle, thence it flows to the left ventricle. Again most of the blood from the left ventricle re-circulates through the lungs When however, the pressure on the left side exceeds that on the right side, some oxygenated blood from the left side will be shunted to the right side There it will be mixed with the venous blood in the right ventricle and will be pumped out through the aorta to the body and returned by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again

In this malformation as in all cases of complete transposition of the great vessels except for those with pulmonary stenosis it is the relative pressure in the two circula tions which regulates the volume of the shunt Hence there is always severe pulmonary hypertension

Clinical diagnosis The patient shows deep persi tent cyanosis Murmurs are not significant The great size of the pulmonary artery and the relatively easy shunting of blood frequently enable a balance to be established and the heart ceases to enlarge The condition may be compatible with life for a number of years The x ray shows slight cardiac enlargement a concave curve at the base of the heart to the left of the sternum, and extremely vascular lung fields The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy



At one week

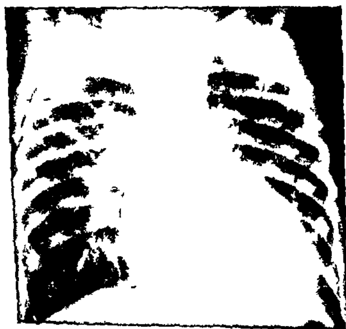


At two weeks

FIGURE X–17 Complete transposition of the great vessels with a dilated pulmonary artery Infant



At one and one quarter years



At two and one-quarter years

FIGURE X–18 Complete transposition of the great vessels with a dilated pulmonary artery Child

dance is the rule Regardless of the presence or absence of a hilar dance, conspicuous vascular shadows indicate a huge pulmonary artery, when this is combined with a concave curve at the base of the heart, as shown in Figure x–19, it is diagnostic of a large posteriorly placed pulmonary artery Hence these findings are almost pathognomonic of a complete transposition of the great vessels combined with dilatation of the pulmonary artery In rare instances the main pulmonary artery is so greatly dilated that it gives the appearance of fullness of the pulmonary conus (see Figure x–20)

In the left anterior oblique position both ventricles are seen to be enlarged and the huge pulmonary artery fills the pulmonary window

In the right anterior-oblique position the left auricle is seen to be of normal size

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram is similar to that in other cases of transposition of the great vessels There is a right axis deviation in the standard leads and evidence of right ventricular hypertrophy in the unipolar precordial leads

SPECIAL TESTS

The circulation time (arm to tongue) is abnormally short

The oxygen saturation of the arterial blood is abnormally low and falls still further with exercise

Cardiac catheterization is usually not necessary for diagnosis but is of aid in the determination of the structure of the auricular septum and whether or not there is a single ventricle When the pulmonary artery over rides the ventricular septum it may be possible to catheterize the pulmonary artery If the pulmonary artery is entered, the oxygen content of the sample of blood in the pulmonary artery will be found to be higher than that in the aorta, indeed, the blood in the pulmonary artery is usually almost fully saturated The pressure in the pulmonary artery is always abnormally high Thus the catheterization findings are closely similar to those of a Taussig Bing malformation Indeed, it is the course which the catheter takes as it enters the pulmonary artery which indicates whether the pulmonary artery occupies its normal position or lies posterior to the aorta

Angiocardiography is almost always disappointing because the dye is dissipated from the right ventricle to both circulations and also because the large size of the pulmonary vessels, combined with the large volume of blood directed to

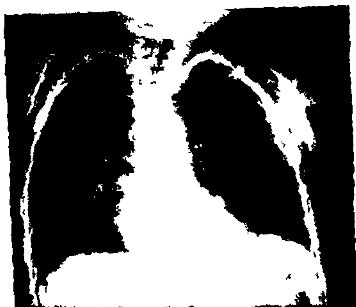


FIGURE X-19 Complete transposition of the great vessels with a greatly dilated pulmonary artery Child

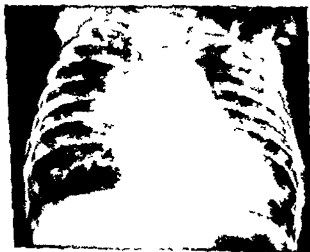


FIGURE X-20 Complete transposition of the great vessels with a greatly dilated pulmonary artery Infant

dance is the rule Regardless of the presence or absence of a hilar dance, conspicuous vascular shadows indicate a huge pulmonary artery, when this is combined with a concave curve at the base of the heart, as shown in Figure X–19, it is diagnostic of a large posteriorly placed pulmonary artery Hence these findings are almost pathognomonic of a complete transposition of the great vessels combined with dilatation of the pulmonary artery In rare instances the main pulmonary artery is so greatly dilated that it gives the appearance of fullness of the pulmonary conus (see Figure X–20)

In the left anterior oblique position both ventricles are seen to be enlarged and the huge pulmonary artery fills the pulmonary window

In the right anterior-oblique position the left auricle is seen to be of normal size

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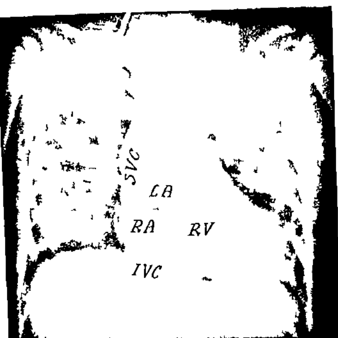
SPECIAL TESTS

The circulation time (arm to tongue) is abnormally short

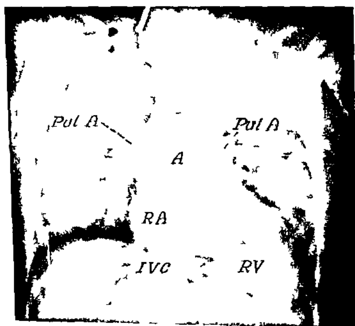
The oxygen saturation of the arterial blood is abnormally low and falls still further with exercise

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Angiocardiography is almost always disappointing because the dye is dissipated from the right ventricle to both circulations and also because the large size of the pulmonary vessels, combined with the large volume of blood directed to



Dye in the superior vena cava, the right auricle and the right ventricle



Dye immediately passes to the aorta and the pulmonary artery

FIGURE X–21 Complete transposition of the great vessels with a posteriorly placed dilated pulmonary artery Child

the pulmonary artery from the left ventricle, causes excessive dilution of th in the lungs (see Figure x–21) Films taken with the patient in the lateral tion, however, confirm the fact that the pulmonary artery arises far poster (see Figure x–22)

DIAGNOSIS

The diagnosis is suggested by the deep, persistent cyanosis and clubbing patient with evidence of a poorly functioning heart and slight enlargemen the liver and congestion in the lungs It is confirmed by the x ray and by flu scopic findings of large, blotchy hilar shadows which may or may not sho conspicuous hilar dance, combined with a concave curve at the base of the h to the left of the sternum which shows that the pulmonary artery is posteri placed

DIFFERENTIAL DIAGNOSIS

The two conditions with which this malformation are most frequently c fused are (1) a single ventricle combined with a complete transposition of great vessels and enormous dilatation of the pulmonary artery and (2) a Ta sig-Bing malformation

A single ventricle combined with complete transposition of the great ves and enormous dilatation of the pulmonary artery may closely resemble the m formation under discussion When the great vessels are completely transposed patient with a single ventricle usually does better than a patient with two ven cles, as there is a better admixture of venous and arterial blood Cardiac cath terization is usually necessary to differentiate the two conditions It is the diff ence in the oxygen content of the blood in the common ventricle in comparis with that in the right auricle which gives the clue to the correct diagnosis Wh there are two ventricles, the increase in the oxygen content of the blood in t right ventricle is comparatively slight, whereas, if there is but a single ventric there is usually an increase of 5 volumes per cent (see Chapter xv)

If there is a defect in the auricular septum, it will cause an increase in th oxygen content of the blood in the right auricle in comparison with that in th superior vena cava Needless to say, this will alter the percentage of increase i the oxygen content of the blood in the ventricle in comparison with that in th right auricle

A Taussig Bing malformation may occasionally be confused with the mal formation under discussion, because in certain instances the contours of the hear are similar In a complete transposition of the great vessels the greatly dilatec

(compare Figures x-22 and xi-6) Functionally the two malformations are closely *similar* but accurate differentiation is important if surgery is contemplated

TREATMENT

Patients with an intact auricular septum can be helped either by a Baffes operation or by the creation of an auricular septal defect, provided that the oxygen content of the blood in the pulmonary *artery is significantly* greater than that in the aorta If, however, the oxygen content of the blood in the aorta and in the pulmonary artery are approximately the same, as in the case of a single ventricle, there is little to gain from operation

PROGNOSIS

The prognosis is usually better when the pulmonary artery is abnormally large than when both great vessels are of normal size A number of patients with the former condition live until late childhood and some live to adult life

During infancy and childhood the increased pressure in the lesser circulation *increases the volume of oxygenated blood shunted into the aorta* Nevertheless, the condition leads to progressive injury to the pulmonary vascular bed and eventually it leads to a reduction in the circulation to the lungs and hence reduces the volume of the effective flow These changes cause the patient to become progressively more incapacitated as he reaches adult life

SUMMARY

Complete transposition of the great vessels combined with enormous dilatation of the pulmonary artery is usually associated with a ventricular septal defect and slight over riding of the pulmonary artery Under these circumstances there is better opportunity for the crossing of the two circulations than in other types of complete transposition of the great vessels Cyanosis becomes apparent at an early age and is of uniform distribution Polycythemia and clubbing usually develop during the first two years of life Respirations are *rapid and shallow* and the infant s chest becomes barrel shaped The liver becomes enlarged The heart is slightly enlarged A gallop rhythm or a pathological third heart sound is audible at the apex

The x ray and fluoroscopic findings are distinctive in that there is evidence of a *huge pulmonary artery* combined with a concave curve at the base of the heart to the left of the sternum

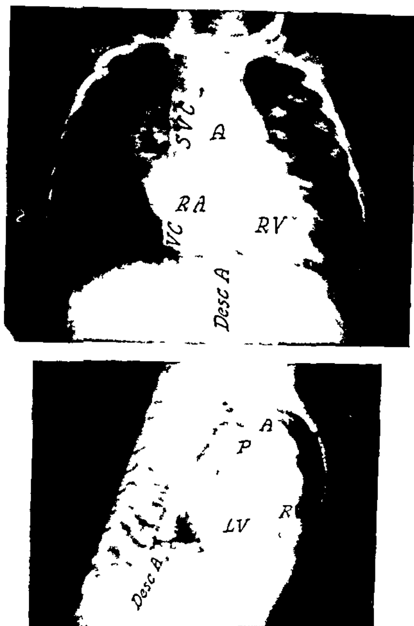


Figure X-22 Complete transposition of the great vessels with an abnormally large pulmonary artery Child

pulmonary artery may cast a shadow to the left of the aorta and thus cause fullness of the pulmonary conus instead of the usual concavity (compare Figures X-20 and XI-4) In the author's experience the vascularity of the lungs is usually more exaggerated when the pulmonary artery arises from the left ventricle Angiocardiography will help to differentiate the two conditions In a Taussig Bing malformation the anterior posterior series of films shows simultaneous opacification of both great vessels as they lie side by side When the great vessels are transposed the pulmonary artery will be seen to lie posterior to the aorta

(compare Figures X-22 and XI-6) Functionally the two malformations are closely similar but accurate differentiation is important if surgery is contemplated

TREATMENT

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The x ray and fluoroscopic findings are distinctive in that there is evidence of a huge pulmonary artery combined with a concave curve at the base of the heart to the left of the sternum

The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy Cardiac catheterization is often necessary to differentiate this malformation from a single ventricle with a complete transposition of the great vessels Angiocardiography is occasionally necessary to differentiate the condition from a Taussig Bing malformation Surgical correction is both difficult and unsatisfactory Nevertheless, a Baffes procedure or the creation of an auricular septal defect is of value if the oxygen content of the blood in the aorta is appreciably lower than that in the pulmonary artery

The prognosis is guarded Although the condition may be compatible with life for ten to twenty years, most persons with this malformation are severely handicapped and die at an early age

C *Complete Transposition of the Great Vessels Combined with Pulmonary Stenosis or Atresia*

Complete transposition of the great vessels may occur in combination with pulmonary stenosis or atresia In both instances the pulmonary obstruction protects the lungs but it also greatly reduces the volume of the pulmonary circulation When there is pulmonary atresia, the pulmonary circulation is so poor that the condition is not long compatible with life

When pulmonary stenosis is combined with a complete transposition of the great vessels, the pressure in the pulmonary artery is low Although the circulation is far more nearly adequate than when there is pulmonary atresia, the reduction in the volume of the pulmonary blood flow is far greater than in other types of complete transposition of the great vessels Indeed, the clinical picture more closely resembles that of a tetralogy of Fallot with a severe pulmonary stenosis than it does other types of complete transposition of the great vessels

NATURE OF THE MALFORMATION

As in most cases of complete transposition of the great vessels, the aorta usually arises from the mid portion of the right ventricle and the pulmonary artery lies directly posterior to it

When there is pulmonary atresia, the only way for the blood to reach the lungs is through the ductus arteriosus or by the collateral circulation The pulmonary blood flow is very meager Little blood is returned to the left auricle, hence only a small volume of blood is pumped out from the left ventricle, the left ventricle remains small, the right ventricle, which pumps the blood to the systemic circulation, becomes hypertrophied (see Figure x-23)

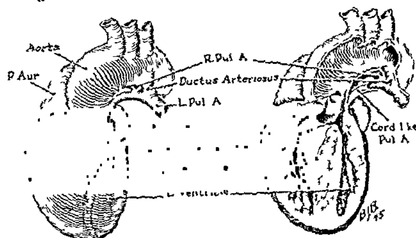


FIGURE X-23 Complete transposition of the great vessels combined with pulmonary atresia and a high ventricular septal defect

When there is pulmonary stenosis, more blood reaches the lungs and more blood is returned to the left auricle and the left ventricle The pulmonary stenosis, however, renders it difficult for the blood to be expelled from the left ventricle, therefore the left ventricle becomes hypertrophied When the pulmonary artery arises from the left ventricle, the pulmonary stenosis is almost always valvular in type, as there is no infundibular chamber from which it can arise The right ventricle pumps the blood to the systemic circulation, hence it, too, is hypertrophied Figure x-24 shows a complete transposition of the great vessels combined with a valvular pulmonary stenosis in which there is a moderately large ventricular septal defect Such is the usual finding in this malformation

In some instances the aorta is further transposed and occupies the position of the normal pulmonary artery Under such circumstances, the ascending aorta arches boldly to the left and turns to the right and may descend on the left or the right of the vertebral column

As in all cases of complete transposition of the great vessels, there is always some additional malformation A ventricular septal defect is common, occasionally there is only an auricular septal defect

COURSE OF THE CIRCULATION

During fetal life the course of the circulation is altered both by the abnormal position of the aorta and by the pulmonary stenosis or atresia Most of the blood in the right auricle flows into the right ventricle and is pumped out through the aorta to the body of the fetus and is returned by the superior vena cava and the

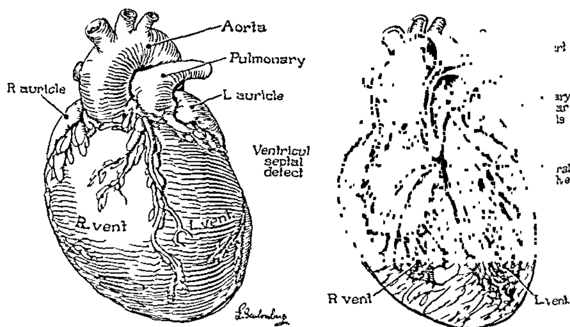


FIGURE X–24 Complete transposition of the great vessels combined with valvular pulmonary stenosis and a ventricular septal defect

inferior vena cava to the right auricle Some of the blood from the right auricle flows into the left auricle and thence to the left ventricle Here the blood encounters difficulty in reaching the aorta, hence the pressure on the left side of the heart tends to rise and this, in turn, tends to lessen the volume of blood which flows from the right auricle to the left The body of the fetus receives adequate circulation from the aorta Even though the pulmonary artery is atretic at its base, *the ductus arteriosus and the main branches of the pulmonary artery* develop normally and the circulation to the lungs is established from the aorta through the ductus arteriosus to the pulmonary artery (see Figure X–25) The fetal circulation is more easily established when there is pulmonary stenosis (see Figure X–26) than when there is pulmonary atresia

After birth the course of the circulation is essentially the same The expansion of the lungs lowers the pulmonary pressure and increases the volume of blood which flows through the ductus arteriosus to the lungs, where it is oxygenated The oxygenated blood is returned to the left auricle The valve covering the foramen ovale tends to close The blood from the left auricle is directed into the left ventricle Inasmuch as no vessel arises from the left ventricle, blood must be pumped from the left ventricle through the septal defect into the aorta The blood which is pumped into the aorta is returned in the normal manner by the superior vena cava and inferior vena cava to the right auricle and thence it

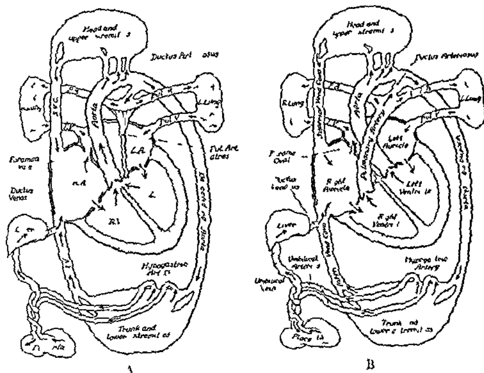


FIGURE X-25 Fetal circulation (A) Complete transposition of the great vessels combined with pulmonary atresia and (B) normal heart

is again pumped out to the systemic circulation The course of the circulation is shown in Diagram X-6

When there is pulmonary stenosis the circulation to the lungs is slightly more adequate as some oxygenated blood from the left ventricle is pumped through the stenosed artery to the lungs through which it circulates and is returned to the left auricle All the blood which is pumped out from the right side of the heart into the aorta is returned by the superior and inferior venae cavae to the right auricle and the right ventricle As in all cases of complete transposition of the great vessels, some crossing of the two circulations must occur There must be either an auricular or a ventricular septal defect or an anomaly of the venous return In the malformation under discussion the pulmonary stenosis renders it difficult for the left ventricle to empty itself The pressure within the left ventricle is increased When the pressure in the left ventricle exceeds that in the right ventricle oxygenated blood is directed to the systemic circulation through the ventricular septal defect When the pressure is greater on the right side of the heart, some venous blood is directed through the defect into the stenosed pulmo-

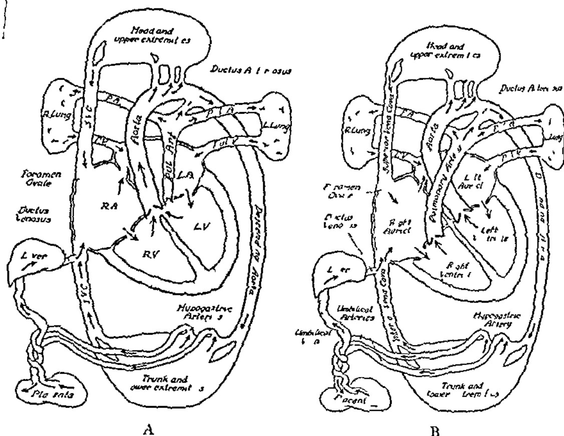


FIGURE X-26 Fetal circulation (A) Complete transposition of the great vessels combined with pulmonary stenosis and (B) normal heart

nary artery and thence to the lungs The course of the circulation is shown in Diagram X-7

When there is an auricular septal defect and no ventricular septal defect, the pressure in the left ventricle will continue to rise until the pressure in the left auricle exceeds that of the right auricle and thereupon a left to-right shunt will be established The volume of the shunt will, however, be small and the effective flow will be extremely low The course of the circulation is shown in Diagram X-8

PHYSIOLOGY OF THE MALFORMATION

Inasmuch as the aorta arises from the right ventricle, the systolic pressure in the right ventricle is the same as that in the systemic circulation Furthermore, since the only way for blood to escape from the left ventricle is through the stenosed pulmonary artery or through the septal defect to the right ventricle, the pressure in the left ventricle is approximately the same as that in the right ven

tricle Indeed, it is the relative pressure in the two ventricles which regulates both the volume and the direction of the shunt The pulmonary stenosis or atresia, however, protects the lungs and the pressure in the pulmonary artery is low

When there is pulmonary atresia, the entire circulation to the lungs is by way of the ductus arteriosus Hence, only if the collateral circulation develops with great rapidity, is the baby able to survive the closure of the ductus arteriosus

When there is pulmonary stenosis, the pulmonary blood flow is greater than when the pulmonary artery is atretic Nevertheless, there is great difficulty in the direction of venous blood to the lungs and of arterial blood to the systemic circulation The effective flow is very low

The difficulty in the ejection of blood from the left ventricle raises the pressure in that chamber Indeed the pressure in the left ventricle must equal or exceed the pressure in the right ventricle in order to direct oxygenated blood to the systemic circulation When the pressure in the left ventricle is lower than that in the right ventricle, some blood from the right ventricle will be pumped into the left ventricle this is the main pathway by which venous blood can be directed to the lungs after the closure of the ductus arteriosus

If the ventricular septum is intact the pressure in the left ventricle will rise still further this in turn will raise the pressure in the left auricle When the pressure in the left auricle exceeds that in the right, a left to-right shunt will be established at the auricular level Over a period of years the collateral circulation develops which aids in the direction of venous blood to the lungs

CLINICAL FINDINGS

During infancy the clinical findings are closely similar to those of a tetralogy of Fallot with severe pulmonary stenosis

Cyanosis is always intense and usually dates from birth It is of uniform distribution

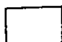
Attacks of paroxysmal dyspnea occur early in life as the ductus arteriosus undergoes obliteration Infants with pulmonary atresia frequently suffer from episodes of loss of consciousness and seldom survive the closure of the ductus arteriosus When, however, there is pulmonary stenosis the condition may be compatible with life for many years

Clubbing develops early and becomes pronounced

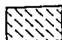
Polycythemia develops rapidly The red blood cell count may reach 11 million cells per cu mm, and the amount of available hemoglobin will be proportionately increased The hematocrit reading may be above 90 per cent Over a

DIAGRAM x–6

Head and upper extremities
Ductus Arteriosus
R Lung
S.V.C.
P.A.
Pul. A.
L Lung
Aorta
P.V.
Pul. V.
L.A.
Remnant of Pul. Art.
R.A.
L.V.
R.V.
Descending Aorta
I.V.C.
Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

Diagram x-6

Complete transposition of the great vessels pulmonary atresia a high ventricular septal defect and a patent ductus arteriosus

In this malformation the aorta arises primarily from the right ventricle and the pulmonary artery is atretic hence it makes no difference whether the pulmonary artery arises from the right or the left ventricle

The blood from the right auricle flows into the right ventricle and is pumped out through the aorta to the body The blood in the systemic circulation is returned in the normal fashion by the superior vena cava and the inferior vena cava to the right auricle and thence to the right ventricle Since there is pulmonary atresia, the only way for the blood to reach the lungs is through the ductus arteriosus to the pulmonary artery The blood which goes to the lungs is returned in the normal manner by the pulmonary veins to the left auricle and thence to the left ventricle. The blood from the left ventricle is pumped out into the aorta Inasmuch as the aorta arises mainly from the right ventricle there is difficulty in the expulsion of blood from the left ventricle If the duration of life is sufficiently long the left ventricle may be hypertrophied, usually the left ventricle is small The right ventricle does the main work it is hypertrophied

Clinical diagnosis The heart is enlarged especially the right ventricle In the anterior posterior position there is no fullness of the pulmonary conus and in the left anterior-oblique position the pulmonary window is wide and clear

In this malformation there is not only difficulty in the direction of the blood to the lungs for oxygenation but because the aorta arises primarily from the right ventricle, there is also difficulty in the direction of the oxygenated blood from the left ventricle into the aorta The body receives mainly venous blood from the right ventricle consequently cyanosis and dyspnea are intense The condition is not long compatible with life

DIAGRAM X–7


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

DIAGRAM X–7

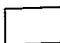
Complete transposition of the great vessels pulmonary stenosis a high ventricular septal defect and a patent foramen ovale

In this malformation there is complete transposition of the great vessels combined with pulmonary stenosis and a high ventricular septal defect The foramen ovale is frequently held open by the high pressure in the right auricle it permits the flow of blood in one direction only, namely, from right to left

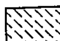
The blood from the right auricle flows into the right ventricle, is pumped out by way of the aorta to the systemic circulation and is returned by the superior and inferior venae cavae to the right auricle and thence to the right ventricle The blood from the left auricle flows into the left ventricle and is pumped out by way of the pulmonary artery to the lungs whence it is returned by the pulmonary veins to the left auricle The only possible way for a crossing of the two circulations is either from the right auricle through the foramen ovale to the left auricle or from either one of the ventricles through the high ventricular septal defect into the aorta or the pulmonary artery As in all cases of complete transposition of the great vessels the blood shunted from one side to the other is returned to the side to which it was shunted it thereby raises the pressure on that side and lowers the pressure on the side from which it was shunted Thus blood shunted from the left ventricle into the aorta is returned to the right auricle and the right ventricle As the pressure in the right ventricle rises blood can be shunted through the ventricular septal defect into the stenosed pulmonary artery Nevertheless, inasmuch as there is pulmonary stenosis little blood reaches the lungs The pressure in the right ventricle rises and sub equently the pressure in the right auricle also rises The valve covering the foramen ovale is forced open and the blood is shunted from the right auricle to the left auricle thence to the left ventricle, and out by way of the pulmonary artery and the aorta Owing to the small size of the pulmonary artery, there is a greater tendency for the blood to be shunted from the left ventricle into the aorta than from the right ventricle into the pulmonary artery There is little tendency for a reversal in the direction of the shunt In contrast to most cases of complete transposition of the great vessels the heart does not undergo rapid progressive enlargement

Clinical diagnosis Cyanosis and clubbing are intense Physical development is retarded Stunting of growth may be extreme The heart is but slightly enlarged The shadow at the base of the heart to the left of the sternum is concave and the second sound in this region is accentuated The murmur if present is systolic in time The x ray shows a contour similar to that of a tetralogy of Fallot but the vascular markings extend nearly to the periphery of the lungs The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy

Diagram X-8


Arterial blood (fully saturated)


Venous and arterial blood. Cyanosis visible


Small admixture of venous blood. No visible cyanosis


Venous blood

Diagram X-8

Complete transposition of the great vessels pulmonary stenosis and an auricular septal defect

The essential feature of this malformation is complete transposition of the great vessels combined with pulmonary stenosis the ventricular septum is intact but there is a gross defect in the auricular septum

The blood from the right auricle flows into the right ventricle and is pumped out through the aorta to the body and returned by the superior vena cava and the inferior vena cava to the right auricle In a similar manner the blood from the left auricle flows into the left ventricle and is pumped out through the pulmonary artery to the lungs and returned by the pulmonary veins to the left auricle With the first breaths of life, the pulmonary pressure drops and thus lowers the pressure on the left side of the heart Hence more blood is directed from the right auricle to the left auricle. This continues until the pressure in the left auricle exceeds that of the right auricle thereupon the direction of the shunt is reversed

The pulmonary stenosis protects the lungs but increases the work of the left ventricle As the pressure in the left ventricle rises the pressure in the left auricle is increased The shunt is at the auricular level but it is the relative pressure in the two ventricles which regulates the volume and direction of the shunt.

Clinical diagnosis Cyanosis and clubbing are intense Stunting of growth is extreme The clinical findings and the cardiac findings are similar to those of a complete transposition of the great vessels with pulmonary stenosis and a ventricular septal defect except that there is more train on the auricles and both the right auricle and left auricle are usually enlarged The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy

period of years the extreme polycythemia leads to a decrease in the blood platelets and to the usual changes in the clotting mechanism of the blood

Respirations are rapid, especially in early infancy As the child grows and compensation improves, the respiratory rate slows but always remains accelerated

Barrel shaped chest deformity is the rule

The liver is usually slightly enlarged but does not pulsate

The pulse pressure is narrow The pulses may be so weak that they are difficult to feel, nevertheless, they are of equal strength in the upper and lower extremities

The blood pressure is low

Stunting of growth is usually extreme In no other malformation is the stunting of growth as extreme as that which occurs in transposition of the great vessels A lad of sixteen years may have the stature of a boy of twelve, or a boy of twelve may be as small as the average child of eight Severe stunting of growth in a cyanotic child should always arouse suspicion of a transposition of the great vessels with pulmonary stenosis

Exercise tolerance is extremely limited The child with a complete transposition of the great vessels and pulmonary stenosis seldom learns to walk before three years of age and frequently not until the age of five or six, even then he can walk only a short distance Many of these children get relief by squatting and develop this habit as soon as they learn to walk

CARDIAC FINDINGS

The heart is usually slightly enlarged

A systolic murmur and a *thrill* over the base of the heart to the left of the sternum are the rule Usually, however, neither the murmur nor the thrill is very intense

The second sound at the base is usually better heard to the left of the sternum than to the right, because the abnormal position of the aorta renders the "pulmonic" second sound louder than the "aortic" second sound There is, however, no reduplication of the second sound

Terminally there may be right sided cardiac failure with engorgement of the liver but congestion in the lungs is minimal

X RAY AND FLUOROSCOPIC FINDINGS

There are two distinct x ray pictures, depending on the position of the aorta

When the aorta arises from the right ventricle but is not rotated abnormally far the contour of the heart is closely similar to that of a tetralogy of Fallot Nevertheless during infancy the heart is usually slightly larger and the vascular markings are more conspicuous and extend further toward the periphery of the lungs than in a tetralogy of Fallot

If the pulmonary stenosis is not too severe and the condition is compatible with life for a number of years as the child grows and the diaphragm descends the contour of the heart comes to resemble that of a tetralogy of Fallot The heart is however slightly enlarged There is a concave curve at the base of the heart to the left of the sternum and there are dense hilar shadows In addition there are many small discrete circular shadows due to the blood vessels seen on end as they course from the posteriorly placed pulmonary artery to the anterior portion of the lung (see Figure x-27) Careful examination of the x ray film will show



FIGURE X 27 Complete transpo t on of the great vessels comb ned w th pulmonary stenos s Adult

that the vascular markings extend nearly to the periphery These findings are more readily seen on the x ray film than upon fluoroscopy Hence it is often the discrepancy between these two observations which gives the clue to the diagnosis

In the left anterior oblique position the pulmonary window is clear, as the posteriorly placed pulmonary artery courses to the lungs at a lower level than when it arises from the right ventricle

When the aorta is rotated so far to the left that it occupies the position of the normal pulmonary artery, there is marked fullness of the pulmonary conus, as shown in Figure x–28 The conspicuous size of the 'pulmonary conus' is in striking contrast to the clarity of the lungs In reality the shadow high up to the left of the sternum is cast by the aorta, the branches of the posteriorly placed pulmonary artery are not visible as they course to the lungs In the left anterior oblique position the pulmonary window is large and clear



FIGURE x–28 Complete transposition of the great vessels combined with pulmonary stenosis Child

The aorta is transposed far to the left

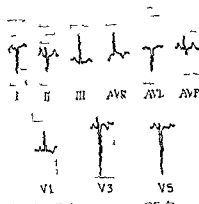


FIGURE X-29 *Complete transposition of the great vessels combined with pulmonary stenosis*

ELECTROCARDIOGRAPHIC FINDINGS

The standard leads show a right axis deviation and the unipolar precordial leads show evidence of right ventricular hypertrophy (see Figure X-29)

SPECIAL TESTS

The condition of an infant with a complete transposition of the great vessels and pulmonary atresia is usually too critical to permit of special studies Hence the following discussion pertains to patients with pulmonary stenosis

The circulation time is abnormally short Two to four seconds from arm to tongue is not unusual

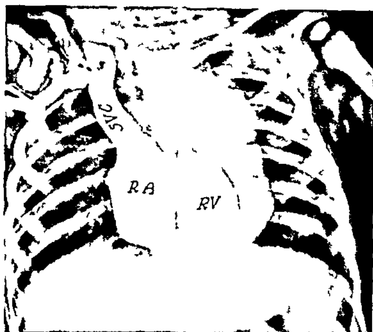
The oxygen saturation of the arterial blood is always reduced and may be extremely low It falls still further with exercise

Cardiac catheterization will usually show that the oxygen content of the blood samples taken from the right auricle, the right ventricle, and the aorta are closely similar It is usually possible to catheterize the aorta but it is manifestly impossible to catheterize the pulmonary artery There may or may not be evidence of an auricular or a ventricular septal defect

Angiocardiography may be of value when the aorta is rotated far to the right, as it will confirm the observation that the vascular shadow at the base of the heart to the left of the sternum is the aorta and not the pulmonary artery (see Figure X-30) Owing to the pulmonary stenosis, no appreciable concentration of dye ever reaches the lungs

DIAGNOSIS

In early infancy the diagnosis is based upon the finding of persistent cyanosis, which appears at an early age and is associated with severe polypnea and fre



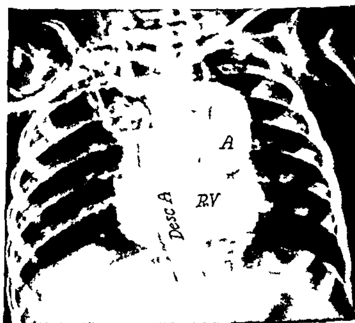


FIGURE X-30 Complete transposition of the great vessels combined with pulmonary stenosis Infant

The aorta rises far to the left and arches to the right

quent attacks of paroxysmal dyspnea, combined with a heart of characteristic contour The concave curve of the cardiac silhouette to the left of the sternum at the base of the heart indicates that the pulmonary artery is diminutive, misplaced, or absent In the left anterior-oblique position the right ventricle is seen to be enlarged The enlargement of the right ventricle and the strength of the pulse indicate that the right ventricle is doing the major work in the maintenance of the circulation The clear pulmonary window indicates that the pulmonary artery is diminutive or absent From these findings it is adduced that the aorta arises mainly from the right ventricle and that the pulmonary artery is markedly stenosed or atretic

In childhood the diagnosis is based upon the finding of a severely incapacitated cyanotic patient with marked stunting of growth who has slight cardiac enlargement and accentuation of the second sound at the base of the heart to the left of the sternum and border line compensation

X ray and fluoroscopic examination aid in the diagnosis The contour of the heart is similar to that of a tetralogy of Fallot but the hilar shadows are dense and the vascular markings extend to the periphery of the lungs When the aorta is rotated far to the left, it occupies the position of the normal pulmonary artery Under such circumstances the aorta arches conspicuously upward to the left of the sternum and the lungs are phenomenally clear

DIFFERENTIAL DIAGNOSIS

The condition is most frequently mistaken for a tetralogy of Fallot, a single ventricle with transposition of the great vessels, or a truncus arteriosus with reduced pulmonary blood flow Occasionally it requires differentiation from a Taussig Bing complex

A tetralogy of Fallot especially one in which there is extreme dextroposition of the aorta, may be extremely difficult to differentiate from a complete transposition of the great vessels with pulmonary stenosis The second sound to the left of the sternum is increased in intensity when the aorta arises from the right ventricle but the same is true when the dextroposition of the aorta is extreme The extension of the vascular markings to the periphery of the lungs is, however, not seen in a tetralogy of Fallot

A single ventricle with pulmonary stenosis may be confused with the malformation under discussion, especially when the great vessels are transposed (see Figure XV-14) Indeed, cardiac catheterization is frequently necessary to differentiate the two conditions

A truncus arteriosus with reduced pulmonary blood flow usually shows a characteristic contour of the heart The heart is large and occupies a transverse position in the chest and the aortic knob is conspicuous Furthermore, the baby, although intensely cyanotic, usually does better than might be anticipated

A Taussig Bing malformation is confused with a complete transposition of the great vessels and pulmonary stenosis only when the aorta arises far to the left In a Taussig Bing malformation there is a full pulmonary conus and the hilar shadows are increased, whereas in the malformation under discussion, the full pulmonary conus is combined with excessively clear lung fields

TREATMENT

When a complete transposition of the great vessels is combined with pulmonary stenosis, the condition may be greatly helped by a systemic pulmonary anastomosis and the creation of an auricular septal defect Although some defect in the auricular or the ventricular septum is essential for life, if only an anastomosis is made, there is great danger that these openings may not be sufficiently large to enable the left side of the heart to expel the increased volume of blood which it receives as a result of the increased circulation to the lungs Therefore, unless a defect of moderate size can be demonstrated, it is wise to combine the anastomotic procedure with the creation of an auricular septal defect Such a defect enables the increased volume of blood which is returned to the left auricle to flow to the right auricle and thence to the right ventricle and out into the aorta From the aorta some blood again flows through the anastomosis to the lungs and is returned to the left auricle Thus the double procedure not only increases the circulation to the lungs but also permits the establishment of a figure of eight circulation Consequently there is hope that this operation may place a constant, not an ever increasing, load upon the heart

A Baffes operation may be of benefit to children with complete transposition of the great vessels and pulmonary stenosis Furthermore, in children it may not be found necessary to relieve the pulmonary stenosis,[14] because the patient can live to childhood only when the pulmonary stenosis is not extreme

PROGNOSIS

Without operation the prognosis is poor Most patients with this malformation are severely incapacitated but may live through childhood Even with operation the prognosis is guarded Although a patient may be symptomatically

benefited by a systemic pulmonary anastomosis combined with the creation of an auricular septal defect, the red blood cell count, the hemoglobin, and the hematocrit values seldom return to normal levels

SUMMARY

Complete transposition of the great vessels may occur in combination with pulmonary atresia but such a malformation is seldom long compatible with life When complete transposition of the great vessels occurs with pulmonary stenosis, the circulation to the lungs is greater but it is still meager, real difficulty is also encountered in the direction of venous blood to the lungs and of oxygenated blood to the systemic circulation

Patients with this malformation are intensely cyanotic Polycythemia develops early Dyspnea is marked Stunting of growth is extreme Exercise tolerance is extremely limited These children squat when tired

The heart may be only slightly enlarged A systolic murmur and a thrill are the rule X ray and fluoroscopic findings are closely similar to those of a tetralogy of Fallot with an extreme degree of pulmonary stenosis, except that the vascular shadows are more pronounced and extend to the periphery of the lungs When the aorta is rotated so far to the left that it occupies the position of the main pulmonary artery it forms a conspicuous arc above the cardiac shadow Consequently there is fullness of the pulmonary conus combined with excessively clear lung fields

The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy Cardiac catheterization shows that the aorta arises mainly or entirely from the right ventricle Angiocardiography will demonstrate that the shadow high up to the left of the sternum is the aorta

The malformation requires differentiation from a tetralogy of Fallot with severe pulmonary stenosis, from a single ventricle with pulmonary stenosis, from a truncus arteriosus with reduced pulmonary blood flow, and occasionally from a Taussig Bing malformation

Patients with this malformation may be benefited by a systemic pulmonary anastomosis combined with the creation of an auricular septal defect and also by a Baffes operation

Most of these patients are severely handicapped Without operation the prognosis is poor Operation improves the circulation but seldom causes a marked reduction in the polycythemia The patient is definitely benefited but still suffers from some limitation of activity

D *Complete Transposition of the Great Vessels Combined with Complete Interruption of the Aortic Arch*

Complete transposition of the great vessels in combination with complete interruption of the aortic arch presents an unusually distinctive clinical syndrome

NATURE OF THE MALFORMATION

The interruption of the aortic arch usually occurs between the left subclavian artery and the point of entrance of the ductus arteriosus When this occurs, the descending aorta becomes continuous with the pulmonary artery through the ductus arteriosus The heart itself is normally formed The ventricular septum is intact (see Figure x-31) The foramen ovale is generally covered by a valve which is not completely sealed

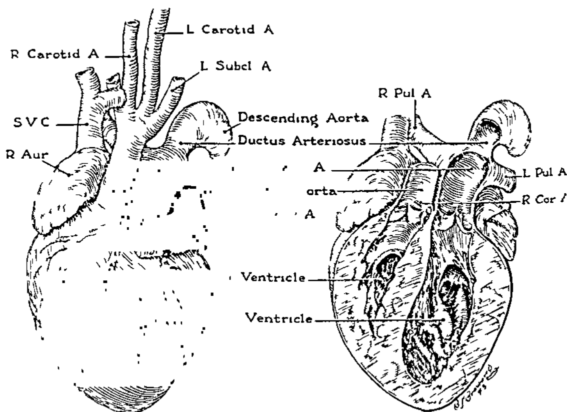


FIGURE x-31 Complete transposition of the great vessels combined with complete interruption of the isthmus of the aorta Infant

The descending aorta is continuous with the pulmonary artery through the ductus arteriosus

COURSE OF THE CIRCULATION

In normal fetal circulation the blood from the pulmonary artery is directed through the ductus arteriosus to the descending aorta and some of the blood from the right auricle is directed through the foramen ovale into the left auricle Essentially the same circulatory mechanism persists when there is complete interruption of the isthmus of the aorta (see Figure XII-2)

When, in addition to the complete interruption of the aortic arch, there is complete transposition of the great vessels, *the blood from the right ventricle* is pumped out through the aorta to the head and the upper extremities, the blood from the left ventricle is pumped out by way of the pulmonary artery to the lungs and also through the ductus arteriosus to the trunk and the lower extremities (see Figure X-32) During fetal life it makes little difference which great vessel pumps the blood to the head and which to the trunk and the lower extremities The malformation places no strain upon the fetal circulation

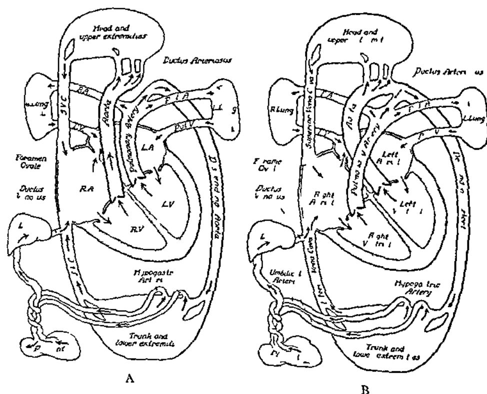


FIGURE X-32 Fetal circulation (A) Complete interruption of the isthmus of the aorta and (B) normal heart

After birth, however, the malformation causes a marked change in the course of the circulation With the expansion of the lungs, the blood in the pulmonary artery is directed to the lungs The consequence is that the blood supply to the trunk and the lower extremities is meager The blood from the lungs is returned in the normal manner to the left auricle With the first breaths of life some blood is also directed from the right auricle through the foramen ovale to the left auricle All the blood from the left auricle flows into the left ventricle and is pumped out through the pulmonary artery to the lungs As soon as the pressure in the pulmonary artery becomes higher than the pressure in the descending aorta, oxygenated blood will again flow from the pulmonary artery through the ductus arteriosus to the descending aorta The blood from the lower extremities is returned by the inferior vena cava to the right auricle The blood from the right auricle passes freely into the right ventricle and is pumped out by way of the aorta to the head and the upper extremities This blood is returned by the superior vena cava to the right auricle In addition, the right auricle receives from the inferior vena cava that portion of the blood from the left ventricle which has been pumped through the ductus arteriosus to the descending aorta and the lower extremities It follows that with each cycle the right auricle receives more blood than was expelled from the right ventricle during the preceding beat Correspondingly, with each successive beat, the left auricle receives less blood than has been pumped out from the left ventricle by way of the transposed pulmonary artery Therefore the pressure in the right auricle will steadily rise and that in the left auricle will fall When the difference in the pressure between the right auricle and the left becomes sufficiently great, the valve which covers the foramen ovale will be forced open and blood will be shunted from the right auricle to the left This is the only mechanism by which the venous blood is directed to the lungs for oxygenation Diagram X-9 illustrates the course of the circulation

PHYSIOLOGY OF THE MALFORMATION

Inasmuch as the aorta arises from the right ventricle, the head and the upper extremities receive venous blood Since the pulmonary artery is continuous with the descending aorta, through the ductus arteriosus, the pressure in the pulmonary artery is the same as that in the descending aorta In other words, the lungs receive blood under systemic pressure Furthermore, the only oxygenated blood which the head and the upper extremities ever receive is that which has been pumped through the ductus arteriosus to the trunk and the lower extremities and returned by the inferior vena cava to the right auricle This blood is mixed

with the even more poorly ...
the head and the upper extremities by the superior vena cava ... [illegible] ...letely deprived of oxygen, this mix
true t[illegible]
ture [illegible]
ger oxygen content

CLINICAL FINDINGS

Cyanosis of the head and the upper extremities is intense

The difference in cyanosis between the upper and the lower extremities is the most significant of all findings Although the pulses are equal in the upper and the lower extremities, the cyanosis is markedly more intense in the upper extremities The difference in cyanosis is due to the fact that the lower extremities receive oxygenated blood from the descending aorta, which is continuous through the patent ductus arteriosus with the pulmonary artery The line of demarcation of the cyanosis lies at the brim of the pelvis The occurrence of the line of demarcation of the cyanosis at this low level indicates that the principal blood supply to the superficial layers of the abdomen is by way of the superficial epigastric arteries the latter arise from the internal mammary arteries, which in turn are branches of the subclavian arteries (see Chapter XXVII) Thus the abdominal wall receives its blood supply from vessels which are given off above the point of interruption of the aorta

Dyspnea and *polypnea* are extreme

The pulse in the lower extremities is of good quality Even though the blood supply is abnormal the strength of the pulse is normal

CARDIAC FINDINGS

The heart enlarges rapidly By four weeks of age it is greatly enlarged A *murmur* may or may not be present If present, it is *systolic* in time Terminally a *gallop rhythm* may develop The liver is enlarged and the lungs are congested

X RAY AND FLUOROSCOPIC FINDINGS

The shape of the heart is significant The shadow at the base of the heart is narrow and the upper margin of the cardiac shadow to the left of the sternum is concave Both ventricles are enlarged The work of the left ventricle is increased because it pumps blood to the lungs as well as to the trunk and the lower extremities whereas the work of the right ventricle is decreased, since it pumps blood only to the head and the upper extremities Consequently the right ventri

Diagram X-9

Head and upper extremities

Ductus Arteriosus

R Lung

L Lung

S.V.C.

P.A.

Aorta

P.V.

Pulmonary Art

L.A.

Descending

Foramen Ovale

R.A.

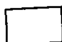
L.V.

R.V.

I.V.C.

Aorta

Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

Diagram X-9

Complete transposition of the great vessels combined with complete interruption of the isthmus of the aorta The descending aorta is continuous with the pulmonary artery through the ductus arteriosus

In this malformation there is in addition to a complete transposition of the great vessels, a complete interruption of the isthmus of the aorta beyond the left subclavian artery, the descending aorta is continuous with the pulmonary artery through the ductus arteriosus Consequently there is always pulmonary hypertension

The blood from the right auricle flows into the right ventricle and is pumped out through the aorta to the head and the upper extremities and is returned by the superior vena cava to the right auricle. The blood in the left auricle flows into the left ventricle and is pumped out into the pulmonary artery The blood which flows to the lungs is returned in the normal manner by the pulmonary veins to the left auricle However some of the blood from the pulmonary artery flows through the ductus arteriosus to the trunk and the lower extremities This blood is returned by the inferior vena cava to the right auricle Thus with each cycle the right auricle receives all the blood pumped out of the right ventricle which is returned by the superior vena cava and some of the blood pumped out of the left ventricle which is returned by the inferior vena cava hence the pressure in the right auricle rises and the pressure in the left auricle falls Consequently the valve covering the foramen ovale is forced open and some blood flows from the right auricle through the foramen ovale to the left auricle This is the only way by which venous blood is directed to the lungs for oxygenation Oxygenated blood is directed to the feet and the lower extremities from the left ventricle through the pulmonary artery by way of the ductus arteriosus The only oxygen which the right side of the heart ever receives is that contained in the venous blood returned by the inferior vena cava Consequently the supply of oxygen to the head and the upper extremities is very meager

Clinical diagnosis The heart is enlarged The shadow at the base of the heart to the left of the sternum is concave The shadow cast by the great vessels is narrow when viewed in the anterior posterior position and increases in width in the left anterior oblique position Cyanosis and dyspnea are intense Furthermore, inasmuch as oxygenated blood is directed from the pulmonary artery through the ductus arteriosus to the descending aorta the lower extremities will be less cyanotic than the upper extremities

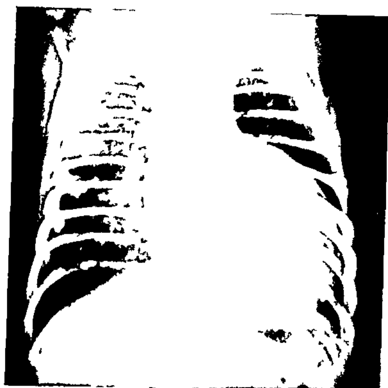


FIGURE X-33 Complete transposition of the great vessels combined with complete interruption of the isthmus of the aorta Infant

cle is not as big as it usually is in most cases of complete transposition of the great vessels and the left ventricle is larger The alteration in the relative size of the two ventricles renders the shape of the heart distinctive The increased size of the left ventricle makes the absence of the pulmonary conus of the right ventricle more conspicuous than it is in most other cases of transposition (compare Figures X-6 and 33) The great size of the left ventricle also causes the apex of the heart to be upturned and the ventricular shadow to extend horizontally toward the axilla In the left anterior oblique position the right ventricle is seen to be enlarged but does not extend as close to the anterior chest wall as in other cases of complete transposition of the great vessels, greater rotation of the infant is required to cause the left ventricle to clear the spinal column

The changes in the width of the shadow at the base of the heart which are characteristic of complete transposition of the great vessels can be demonstrated by fluoroscopic examination In the anterior posterior position the aortic shadow is narrow, upon rotation of the infant into the left anterior oblique position, this shadow increases in width

In addition, rhythmic changes in the size of the right auricle may be seen These are due to the fact that the pressure in the right auricle steadily rises and

that in the left auricle falls until the difference in pressure between the two auricles is sufficient to force open the valve which covers the foramen ovale Thereupon blood will flow from the right auricle to the left auricle and the right auricle will collapse

ELECTROCARDIOGRAPHIC FINDINGS

The standard leads usually show a right axis deviation and the unipolar pre cordial leads should show evidence of greater hypertrophy of the left ventricle than occurs in other cases of complete transposition of the great vessels

DIAGNOSIS

The diagnosis is based upon the finding of an infant with intense cyanosis and severe dyspnea whose feet and lower extremities are of almost normal color The heart is greatly enlarged In the anterior posterior x ray film there is absence of the fullness of the pulmonary conus, a sharp concave curve to the left of the sternum, and a narrow aortic shadow In the left anterior-oblique position the shadow at the base of the heart is increased in width and both ventricles are seen to be enormously enlarged, the right ventricle, however, is slightly smaller and the left ventricle is even larger than is usual in a complete transposition of the great vessels

DIFFERENTIAL DIAGNOSIS

The condition is to be differentiated (1) from a complete transposition of the great vessels with a patent ductus arteriosus the foramen ovale covered by a valve, and a normal aortic arch and (2) from complete interruption of the isth mus of the aorta without complete transposition of the great vessels

Complete transposition of the great vessels with a patent ductus arteriosus and the foramen ovale covered by a valve which is not completely sealed is physi ologically closely similar to the malformation under discussion The contours of the heart in the anterior posterior position are identical (compare Figures x–10 and 33) The two conditions can be differentiated by the difference in the in tensity of the cyanosis between the upper and the lower extremities in the mal formation under discussion the difference is readily apparent, whereas when the aorta is intact it may be so slight that it is difficult to determine with certainty

Complete interruption of the isthmus of the aorta without a transposition of the great vessels can readily be differentiated from the malformation under dis- cussion by the distribution of the cyanosis When the great vessels *occupy their*

normal position, only the feet and the lower extremities are cyanotic, whereas when the great vessels are transposed, the cyanosis is limited to the head, the upper extremities, and the abdominal wall In both instances the line of demarcation of the cyanosis lies at the brim of the pelvis When the cyanosis is limited to the feet, it may be completely overlooked or thought to be due to cold Furthermore, dyspnea is absent

PROGNOSIS

The prognosis is extremely poor The oxygen supply to the head and the upper extremities is so meager that the malformation is compatible with life for only a few weeks

SUMMARY

The diagnostic features of this malformation are (1) intense cyanosis and severe dyspnea, (2) the features which are pathognomonic of a complete transposition of the great vessels the narrow aortic shadow in the anterior posterior position and the wide aortic shadow in the left anterior-oblique position, combined with enlargement of both ventricles and the absence of the fullness of the normal pulmonary conus of the right ventricle, (3) the features which are characteristic of complete absence of the aortic arch between the left subclavian artery and the point of entrance of the ductus arteriosus although the pulses are of equal strength in both extremities, there is a difference in the intensity of cyanosis between the upper and lower extremities, when this occurs in combination with a complete transposition of the great vessels, the cyanosis of the upper extremities is more intense than that of the lower extremities, (4) rhythmic changes in the size of the right auricle indicative of a foramen ovale which is at times functionally open and at times functionally closed

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CHAPTER XI

THE TAUSSIG-BING MALFORMATION

A TRANSPOSITION of the aorta combined with a pulmonary artery which arises from the right ventricle and partially over rides the ventricular septum was listed by Pernkopf in 1926[1] as one possible combination of anomalies in a transposition of the great vessels It was not, however, until 1948 that Taussig and Bing[2] showed that this malformation presented a distinctive clinical syndrome

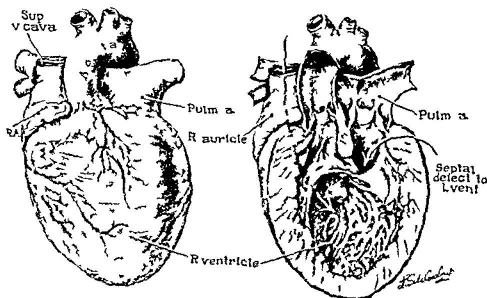
This malformation is relatively common among children who suffer from persistent cyanosis because the structure of the heart permits a fair degree of the crossing of the two circulations and does not lead to progressive cardiac enlargement Therefore the condition is compatible with relative longevity Failure to differentiate this syndrome as a specific entity occurred partly because the anomaly is compatible with relative longevity and partly because both clinicians and pathologists confused it with an Eisenmenger complex The two conditions are, however, quite different In the Eisenmenger complex the aorta arises mainly from the left ventricle and partially over rides the ventricular septum and consequently it receives blood from both ventricles, the pulmonary artery arises entirely from the right ventricle In contrast to this, in the Taussig Bing malformation the aorta is transposed and arises entirely from the right ventricle, the pulmonary artery also arises from the right ventricle but partially over rides the ventricular septum, consequently it is the pulmonary artery that receives blood from both ventricles

The Taussig Bing malformation is also closely similar to the condition in which both the aorta and the pulmonary artery arise from the right ventricle (see Chapter VII) In such an anomaly there is always a ventricular septal defect to enable the blood to escape from the left ventricle Abbott[3] warned that this latter malformation should not be confused with an Eisenmenger complex The defect frequently lies at the base of the pulmonary artery, which, however, does not over ride the opening When the pulmonary artery over rides the ventricular septum the condition is more readily compatible with life than when both great vessels arise from the right ventricle Consequently such is the usual nature of the malformation among those who live until childhood or early adult life

NATURE OF THE MALFORMATION

The outstanding features of this malformation are the aorta is transposed and arises entirely from the right ventricle the pulmonary artery arises mainly from the right ventricle but partially over rides the ventricular septum and consequently also receives blood from the left ventricle there is a high ventricular septal defect and there is right ventricular hypertrophy

In this malformation the transposed aorta lies to the right of the pulmonary artery as shown in Figure XI 1 The aorta is usually smaller than the pulmonary artery the aorta however may be normal in size and the pulmonary artery is frequently abnormally large When the pulmonary artery over rides the ventricular septum the condition is described as levoposition of the pulmonary artery to designate the counterpart of dextroposition of the aorta Dextroposition of the aorta means that although the aorta arises primarily from the left ventricle, it is displaced so that the aortic orifice partially over rides the ventricular septum and consequently the aorta receives blood directly from the right ventricle Levo-



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ventricle is also pumped into the pulmonary artery (see Figure XI-3) Thus the pulmonary artery receives blood from both ventricles Consequently the volume of blood which flows through the pulmonary artery during fetal life is greater than normal This may be the explanation of the abnormally large size of the pulmonary artery

After birth all the blood from the right auricle flows into the right ventricle Part of the blood from the right ventricle is pumped out by way of the aorta to the systemic circulation and part is pumped out by way of the pulmonary artery to the lungs All the blood which goes to the lungs is returned by the pulmonary veins to the left auricle Thence it flows into the left ventricle Most of the blood from the left ventricle is pumped directly into the pulmonary artery, which over rides the ventricular septum and arises in part from the left ventricle This oxy genated blood re-circulates through the lungs and is returned to the left auricle and the left ventricle Inasmuch as the aorta arises from the right ventricle, the only oxygenated blood which reaches the aorta is that which is pumped through

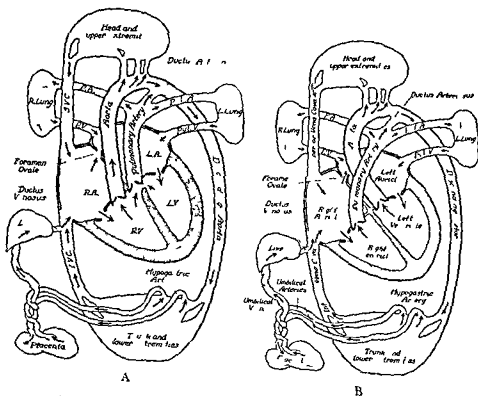


FIGURE XI-3 Fetal circulation (A) Taussig Bing malformation and (B) normal heart

position of the pulmonary artery means that the pulmonary artery, although it arises mainly from the right ventricle, partially over rides the ventricular septum and thereby receives blood from both ventricles, as shown in the cross section in Figure VI-2 A high ventricular septal defect is inevitable either with dextroposition of the aorta or with levoposition of the pulmonary artery In the malformation under discussion the abnormal position of the pulmonary artery causes the defect to lie beneath that vessel Inasmuch as the aorta arises entirely from the right ventricle and the pulmonary artery also arises mainly from the right ventricle, there is right ventricular hypertrophy

COURSE OF THE CIRCULATION

During fetal life the right ventricle does the main work Most of the blood from the right auricle flows into the right ventricle and from there is pumped out both into the aorta and into the pulmonary artery The blood in the aorta is directed to the systemic circulation and is returned in the normal manner to the right auricle The blood in the pulmonary artery flows to the lungs and through the ductus arteriosus to the descending aorta The blood which flows to the trunk and the lower extremities is returned in the normal manner to the right auricle and that which circulates through the lungs is returned in the normal fashion to the left auricle and thence to the left ventricle Inasmuch as the aorta arises entirely from the right ventricle and the pulmonary artery arises partially from the left ventricle, the blood from the left ventricle is pumped out into the pulmonary artery As previously mentioned, part of the blood from the right

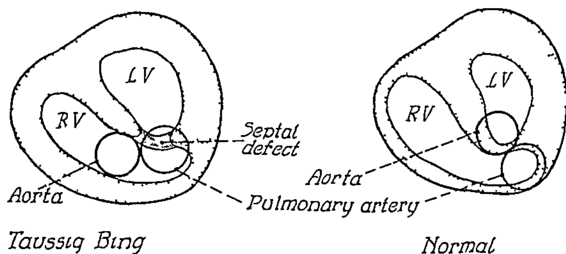


FIGURE VI-2 Taussig Bing malformation and normal heart

between the gradual expansion of the lungs and the pulmonary vascular changes Eventually the pulmonary resistance becomes so high that the right-to-left shunt is increased and the oxygen saturation of the arterial blood is further decreased The collateral circulation develops, polycythemia increases still further and may become extreme

Polypnea is a common complaint Infants with this malformation rarely have attacks of paroxysmal dyspnea such as so frequently occur in infants with pulmonary stenosis or atresia, and they do not suffer from episodes of loss of consciousness such as are common in infants with a complete transposition of both great vessels

The exercise tolerance is limited Nevertheless, the incapacity is not as great as might be expected with a transposed aorta Although the child may tend to squat when he first starts to walk, he soon overcomes the habit He can frequently walk a considerable distance at a slow pace

Stunting of growth may occur *Weight gain* is slow and often the child is very thin

The blood pressure is difficult to obtain and the *pulse pressure* is narrow but of equal strength in the upper and the lower extremities

CARDIAC FINDINGS

The heart is but slightly enlarged It may or may not be possible to detect the fullness of the pulmonary conus by percussion The *second sound* over the pulmonary area is accentuated On palpation there is a *systolic thrill* and on auscultation a *systolic murmur* is audible over the precordium The murmur is usually neither very loud nor rasping

X RAY AND FLUOROSCOPIC FINDINGS

The heart is virtually of normal size The pulmonary conus is full and the hilar markings are increased (see Figure XI–4) In early childhood the contour of the heart may closely resemble that of an uncomplicated patent ductus arteriosus or an auricular septal defect In older children and young adults the *contour* of the heart is closely similar to that of an Eisenmenger complex

The left anterior-oblique position is the most advantageous for the study of the aorta and the pulmonary artery In this view the aorta appears small and the large pulmonary artery arches posteriorly below the aorta

The right anterior-oblique position shows the relation of the pulmonary artery to the esophagus When the pulmonary artery is abnormally large, it may

the septal defect into the right ventricle In the right ventricle the oxygenated blood from the left ventricle mixes with the venous blood from the right auricle, this mixture of oxygenated and venous blood is pumped out by way of the aorta to the systemic circulation and is returned in the normal manner by the superior and inferior venae cavae to the right auricle There the cycle starts again The course of the circulation is shown in Diagram XI-1

PHYSIOLOGY OF THE MALFORMATION

Inasmuch as the aorta arises from the right ventricle, the right ventricle takes over the function of the left ventricle, the pressure in the right ventricle is that which is normal for the left ventricle Consequently the pulmonary artery, which arises from the right ventricle, receives blood under systemic pressure, hence there is always pulmonary hypertension

The pressure in the left ventricle is also elevated, as at best it must expel blood against systemic pressure, in addition, there is difficulty in the ejection of the large volume of blood returned to the left ventricle, for this reason the pressure in the left ventricle may exceed that in the right ventricle This in turn further raises the pressure in the pulmonary artery Consequently the pressure in the pulmonary artery may exceed the systemic pressure Since the oxygenated blood returned to the left ventricle is directed into the pulmonary artery, the oxygen saturation of the blood in the pulmonary artery is higher than that in the aorta

The high pressure with which the blood is ejected into the lungs sets up the usual series of changes in the pulmonary vascular bed As the pulmonary resistance rises, less blood reaches the lungs for oxygenation and more blood is shunted into the aorta As the years go by, these changes gradually lead to a further decrease in the oxygen saturation of the arterial blood and to ever increasing polycythemia Eventually the condition becomes incompatible with life

CLINICAL FINDINGS

The outstanding clinical findings are persistent cyanosis and clubbing of the extremities

Cyanosis dates from birth and is of uniform distribution *Clubbing* of the extremities develops at an early age There is usually dilatation of all the capillaries of the body and suffusion of the conjunctivae

Polycythemia is the rule The red blood cell count, the amount of available hemoglobin, and the hematocrit reading are abnormally high During childhood the hemoconcentration remains relatively constant, as there is a balance

Diagram XI-1

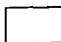
Taussig Bing malformation

In this malformation the aorta is transposed and arises from the right ventricle The pulmonary artery arises mainly from the right ventricle but slightly over rides the ventricular septum and receives blood from both ventricles It follows that the ventricular septal defect is an integral part of the malformation The defect lies beneath the pulmonary artery Since the aorta arises from the right ventricle and the pulmonary artery also arises in part from the right ventricle there is pulmonary hypertension

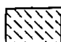
The blood from the right auricle flows into the right ventricle Inasmuch as the aorta arises from the right ventricle part of the blood from the right ventricle is pumped directly into the aorta and part is pumped into the pulmonary artery The blood pumped into the pulmonary artery flows to the lungs, where it is oxygenated and is returned to the left auricle in the normal manner All the blood in the left auricle flows into the left ventricle The blood in the left ventricle is pumped out into the pulmonary artery and through the septal defect into the right ventricle Inasmuch as the pulmonary artery over rides the ventricular septum it receives a mixture of oxygenated blood from the left ventricle and venous blood from the right ventricle. This mixture of oxygenated and venous blood circulates through the lungs and is returned to the left side of the heart The only oxygenated blood which the aorta receives is that which is pumped from the left ventricle through the septal defect into the right ventricle. Thus mixture of oxygenated and venous blood which has a lower oxygen content than that in the pulmonary artery is pumped into the aorta and circulates through the body and is returned in the normal manner by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again

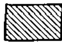
Clinical diagnosis Cyanosis dates from birth and is persistent Polycythemia and clubbing of the extremities develop at an early age The patient s exercise tolerance is limited but he does not squat when tired The heart is but slightly enlarged The contour of the heart is similar to that seen in an Eisenmenger complex in that there is fullness of the pulmonary conus Pulsations in the hilar shadows develop at an early age The fact that both the aorta and the pulmonary artery arise from the right ventricle increases the work of that chamber The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy

Diagram XI–1


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

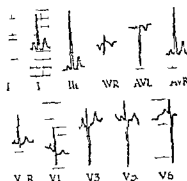


FIGURE XI-5 Taussig Bing malformation (Case XI-1) Child

nto the right diverts the catheter from the pulmonary orifice If the catheter does not enter the pulmonary artery, the findings are similar to those in a tetralogy of Fallot If the pulmonary artery is catheterized, *the pressure in the* pulmonary artery is as high or higher than that in the aorta and the oxygen content of the blood in the pulmonary artery is higher than that in the aorta

Angiocardiography reveals early filling of the aorta and slight opacification in the region of the pulmonary conus The dye may be dissipated so rapidly through the large pulmonary artery that little of the radio-opaque material is visible in the lungs (see Figure XI-6)

The procedure, however, is not without danger because the rapid injection of the dye increases the pressure in the right side of the heart and consequently may block the flow of blood from the left ventricle to the right ventricle and thereby may decrease the supply of oxygen to the body Consequently, not only is a large amount of contrast media poured into the aorta, but the patient is simultaneously deprived of oxygen The procedure has been known to cause sudden death Usually, however, the oxygen saturation of the arterial blood is sufficiently high so that the momentary decrease in the oxygen content of the blood is not dangerous for the patient Nevertheless, caution should be exercised, especially as angiocardiography is seldom necessary for the diagnosis of this malformation

DIAGNOSIS

The diagnosis is based upon the contour of the heart in the x ray and the finding of persistent cyanosis which dates from birth The heart is but slightly enlarged and the x ray shows fullness of the pulmonary conus and increased hilar shadows fluoroscopy usually reveals expansile pulsations in the hilar vessels The electrocardiogram shows a right axis deviation and evidence of right ven



FIGURE XI-4 Taussig Bing malformation (Case XI-1) Child

cause backward displacement of the esophagus below the level of the aortic arch

Fluoroscopic examination usually reveals expansile pulsations of the hilar shadows, these become increasingly conspicuous with age

ELECTROCARDIOGRAPHIC FINDINGS

The standard leads show a right axis deviation and the unipolar precordial leads show evidence of right ventricular hypertrophy (see Figure XI-5)

SPECIAL TESTS

The circulation time (arm to tongue) is abnormally short

The oxygen saturation of the arterial blood is abnormally low Upon exercise there is a gradual further fall in the oxygen content of the arterial blood

Cardiac catheterization reveals a high pressure in the right ventricle The oxygen content of the blood in the right ventricle is higher than that in the right auricle It is easy to pass the catheter into the aorta In some instances it is possible to catheterize the aorta and the pulmonary artery with equal ease In other instances difficulty may be encountered in catheterization of the pulmonary artery because the stream of blood which is pumped from the left ventricle

tricular hypertrophy The diagnosis is confirmed by catheterization when the aorta and the pulmonary artery are entered with equal ease

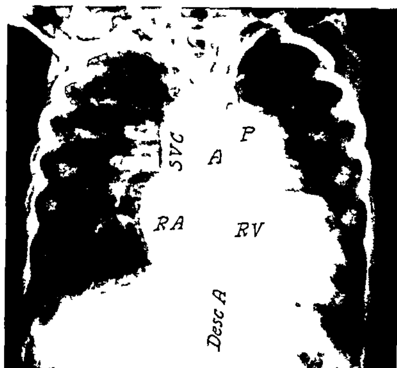
DIFFERENTIAL DIAGNOSIS

The malformation is most commonly confused with an Eisenmenger complex and occasionally with a complete transposition of the great vessels and a dilated pulmonary artery, it may also be confused with a cor pulmonale, that is, with primary pulmonary hypertension

The Eisenmenger complex produces a clinical syndrome which, in older patients, is closely similar to that of the Taussig Bing malformation In both malformations the patients show persistent cyanosis, in neither does the patient squat The contours of the heart and the x ray and fluoroscopic findings are similar both malformations cause fullness of the pulmonary conus and a hilar dance Both show a right axis deviation and evidence of right ventricular hypertrophy Both have a short circulation time The salient difference between the two malformations is that in the Taussig Bing malformation cyanosis dates from birth, whereas in the Eisenmenger complex cyanosis usually develops at or about the time of puberty Hence, if the clinical picture is that of an Eisenmenger complex except that cyanosis dates from birth, the malformation is probably that of the Taussig Bing type

Complete transposition of the great vessels with a dilated pulmonary artery which over rides the ventricular septum is functionally similar to a Taussig Bing malformation [4] When the great vessels are completely transposed, the occurrence of a concave curve at the base of the heart and large blotchy hilar shadows clearly differentiates it from a Taussig Bing malformation Occasionally the main pulmonary artery is so greatly dilated that it is visible to the left of the sternum and gives the appearance of a normally placed dilated pulmonary artery Under such circumstances a complete transposition of the great vessels may closely simulate a Taussig Bing malformation Angiocardiography or observation of the course of the catheter as it enters the pulmonary artery may be necessary to differentiate the two conditions

A partial anomaly of the venous return combined with a Taussig Bing malformation has been reported by Schultz [5] Such an anomaly is of benefit to the patient, as the entrance of some of the pulmonary veins into the right auricle raises the oxygen saturation of the blood in the right auricle and in the right ventricle and increases the volume of oxygenated blood directed into the aorta (see Diagram XI-2) Hence it lessens the patient's incapacity and renders the



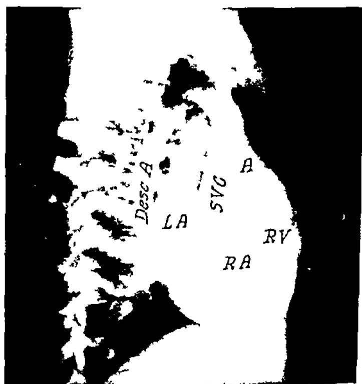


Figure XI–6 Taussig Bing malformation (Case XI–1) Child

Anterior posterior view shows simultaneous opacification of the aorta and the pulmonary artery In the lateral view the pulmonary artery is concealed by the aorta

tricular hypertrophy The diagnosis is confirmed by catheterization when the aorta and the pulmonary artery are entered with equal ease

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Diagram XI–2

Head and upper extremities

R Lung

S.V.C.

P.A.

Aorta

Pulmonary Artery

P.A.

L Lung

PV

L A

PV

R.A.

L V

R.V.

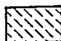
Descending Aorta

I.V.C.

Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis

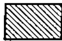

Venous blood

DIAGRAM XI–2

Taussig Bing malformation combined with a partial anomaly of the pulmonary venous return

In this instance a Taussig Bing malformation is combined with the anomalous return of the right pulmonary veins into the right auricle

The blood from the right auricle flows into the right ventricle Since the aorta arises entirely from the right ventricle and the pulmonary artery arises partly from the right ventricle the blood from the right ventricle is pumped out into both the aorta and the pulmonary artery hence there is pulmonary hypertension The blood which goes to the lungs is oxygenated Part of the oxygenated blood from the lungs is returned to the right auricle and the rest is returned to the left auricle thence it flows to the left ventricle The blood in the left ventricle is pumped out into the pulmonary artery and through the septal defect into the right ventricle and thence into the aorta All the blood which is directed into the aorta is returned by the superior vena cava and the inferior vena cava to the right auricle There the venous blood is mixed with the fully oxygenated blood which is returned by the right pulmonary veins to the right auricle This mixture of venous and arterial blood flows into the right ventricle. There the cycle starts again

Clinical diagnosis The diagnosis is based upon the finding of a slightly cyanotic patient with the cardiac findings of a Taussig Bing malformation Cyanosis is minimal because part of the blood which is oxygenated in the lungs is returned by the anomalous vein to the right auricle and thence to the right ventricle thereby the volume of the oxygenated blood directed to the systemic circulation is increased

condition more compatible with life. This combination of anomalies should be considered when the physical findings are similar to those in a Taussig-Bing malformation but cyanosis is minimal.

A gross defect in the auricular septum is also beneficial to the patient. Indeed, such a defect does what the surgeon attempts to do when he creates an auricular defect (see below).

Cor pulmonale and primary pulmonary hypertension in older patients may resemble the Taussig-Bing malformation. However, a patient with a cor pulmonale or primary pulmonary hypertension does not usually show cyanosis at birth. Indeed, the development of cyanosis during adolescence or in early adult life *is* characteristic of a cor pulmonale. Moreover, the severity of the dyspnea is out of proportion to the intensity of the cyanosis. Furthermore, the systemic circulation time is normal or prolonged in patients with primary pulmonary hypertension, whereas in the malformation under discussion the arm-to-tongue circulation time is abnormally short. Either cardiac catheterization or angiocardiography will clearly differentiate the two malformations, as the aorta arises from the left ventricle in a cor pulmonale and from the right ventricle in a Taussig-Bing malformation.

TREATMENT

Surgery offers some hope to patients with a Taussig-Bing malformation. The creation of an auricular septal defect aids in the direction of oxygenated blood to the systemic circulation, and thereby lessens the incapacity of the patient, but it does nothing to alter the pulmonary hypertension.

A Baffes operation[6] is also of real benefit to a patient with a Taussig-Bing malformation, as it aids in the direction of oxygenated blood to the aorta and of venous blood to the pulmonary artery. Neither the Baffes operation nor the creation of an auricular septal defect reduces the pressure in the pulmonary artery. Unfortunately the pulmonary hypertension will eventually cause difficulty.

Medical treatment is the same as for other cyanotic children. Intravenous fluid should always be given extremely slowly, because any procedure which increases the pressure in the right auricle and the right ventricle will lessen the volume of oxygenated blood which is shunted from the left ventricle to the right ventricle and consequently it will reduce the supply of oxygen to the body.

PROGNOSIS

The prognosis varies with the size of the aorta, with the size and position of the pulmonary artery, and with the size of the septal defect. Unless the aorta is

markedly hypoplastic or the septal defect is so small that little oxygenated blood can reach the aorta, the patient generally lives to adult life Nevertheless, the long-continued pulmonary hypertension leads to secondary changes in the lungs, which permit less blood to flow through the pulmonary artery to the lungs, and hence a greater volume of venous blood is directed into the aorta The condition eventually becomes incompatible with life

SUMMARY

The anatomical features of this malformation are a transposed aorta, a large pulmonary artery which arises from the right ventricle and slightly over rides the ventricular septum, a high ventricular septal defect, and right ventricular hypertrophy

The outstanding clinical features are cyanosis which dates from birth, polycythemia, and clubbing of the extremities, combined with a heart which is not greatly enlarged The x ray contour of the heart is similar to that of an Eisenmenger complex because of the fullness of the pulmonary conus, the increased hilar shadows, and the fluoroscopic evidence of a hilar dance The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy The oxygen saturation of the arterial blood is abnormally low The circulation time is abnormally short

Cardiac catheterization shows a high pressure in the right ventricle and the oxygen content of the blood in the right ventricle is higher than that in the right auricle It may be possible to catheterize both the aorta and the pulmonary artery with equal ease if so the diagnosis is readily established by the finding that both the oxygen content of the blood in the pulmonary artery and the pressure in the pulmonary artery are as high or higher than those in the aorta

Angiocardiography, although not usually necessary for diagnosis, may be of value in the rare case in which the contour of the heart in a Taussig Bing malformation simulates that of a complete transposition of the great vessels with a dilated pulmonary artery

The diagnosis is based upon the demonstration of a heart with a contour and fluoroscopic findings similar to those of an Eisenmenger complex in a patient with a history of cyanosis which dates from birth The diagnosis is substantiated by cardiac catheterization provided both the aorta and the pulmonary artery are catheterized and pressures are measured The patient s exercise tolerance is limited, nevertheless, relative longevity is the rule

If the patient is extremely incapacitated, the condition may be ameliorated by the creation of an auricular septal defect or by a Baffes operation Such opera

condition more compatible with life This combination of anomalies should be considered when the physical findings are similar to those in a Taussig Bing malformation but cyanosis is minimal

A gross defect in the auricular septum is also beneficial to the patient Indeed, such a defect does what the surgeon attempts to do when he creates an auricular defect (see below)

Cor pulmonale and primary pulmonary hypertension in older patients may resemble the Taussig Bing malformation However, a patient with a cor pulmonale or primary pulmonary hypertension does not usually show cyanosis at birth Indeed, the development of cyanosis during adolescence or in early adult life is characteristic of a cor pulmonale Moreover, the severity of the dyspnea is out of proportion to the intensity of the cyanosis Furthermore, the systemic circulation time is normal or prolonged in patients with primary pulmonary hypertension, whereas in the malformation under discussion the arm to-tongue circulation time is abnormally short Either cardiac catheterization or angiocardiography will clearly differentiate the two malformations, as the aorta arises from the left ventricle in a cor pulmonale and from the right ventricle in a Taussig Bing malformation

TREATMENT

Surgery offers some hope to patients with a Taussig Bing malformation The creation of an auricular septal defect aids in the direction of oxygenated blood to the systemic circulation, and thereby lessens the incapacity of the patient, but it does nothing to alter the pulmonary hypertension

A Baffes operation[6] is also of real benefit to a patient with a Taussig Bing malformation, as it aids in the direction of oxygenated blood to the aorta and of venous blood to the pulmonary artery Neither the Baffes operation nor the creation of an auricular septal defect reduces the pressure in the pulmonary artery Unfortunately the pulmonary hypertension will eventually cause difficulty

Medical treatment is the same as for other cyanotic children Intravenous fluid should always be given extremely slowly because any procedure which increases the pressure in the right auricle and the right ventricle will lessen the volume of oxygenated blood which is shunted from the left ventricle to the right ventricle and consequently it will reduce the supply of oxygen to the body

PROGNOSIS

The prognosis varies with the size of the aorta, with the size and position of the pulmonary artery, and with the size of the septal defect Unless the aorta is

per cent The oxygen capacity was 20.8 volumes per cent The pressure in the right ventricle was 42/19 mm of mercury and that in the pulmonary artery was 57/45 mm of mercury

Angiocardiography was performed by the technique customary in 1947 This necessitated two injections in order to obtain films in two planes Unfortunately there was a mechanical failure at the time of the second injection and therefore a third injection* was given fifteen minutes later Three minutes thereafter the child sat bolt upright and the heart stopped All efforts at resuscitation failed

The angiocardiograms showed that the dye entered the right auricle and then the right ventricle immediately thereafter the aorta was promptly opacified Little dye was seen in the pulmonary artery or the lungs In the final series of films taken in the lateral position the aorta appeared to arise from the anterior portion of the right ventricle but in this series too the dye could not be traced into the lungs or to the left side of the heart (see Figure XI–6)

Final clinical diagnosis A transposition of the great vessels was indicated by the physiological studies and angiocardiograms x ray and fluoroscopic studies, however, indicated that the pulmonary artery arose in its normal position

Autopsy No 31039 (performed by Dr Edmund Novak) The chief interest centered about the heart It weighed 180 gm. The right auricle was not greatly enlarged The superior vena cava and the inferior vena cava opened into it in the normal fashion The foramen ovale was completely covered by a valve but the margin of the valve showed a probe patency for a distance of 1 0 cm The tricuspid valve which was slightly thickened opened into the right ventricle That chamber was tremendously hypertrophied its wall measured 1 5 cm in thickness The pulmonary artery arose approximately in its normal position The aorta was transposed it arose entirely *from* the right ventricle The aortic orifice lay adjacent to both the pulmonary orifice and the ventricular septum as shown in Figures XI–1 and 2 The aortic valve had three cusps and the coronary arteries were given off from the aorta in the normal manner The aortic ring measured 3 5 cm in circumference The aorta and its branches appeared to be normal The maximum circumference of the ascending aorta was 4.0 cm At the base of the ventricular septum the septal wall was defective for a distance of 1.2 cm and in this region the septal wall was depressed downward approximately 0 6 cm The superior portion of the ventricular septum deviated to the right to such an extent that the pulmonary orifice over rode the septal defect by a few millimeters From the upper margin of the ventricular septum close to the defect a *muscular ridge* extended forward to the outer wall of the right ventricle This ridge separated the aorta from the pulmonary artery Consequently the aorta arose entirely from the right ventricle and only the pulmonary orifice overlay the ventricular septum Thus the

* A warning to prevent repetition of this serious mistake is given on p 235

tions, however, do not relieve the pulmonary hypertension Hence the prognosis remains guarded

Illustrative Case

CASE XI-I P A W (Harriet Lane Home, No A-60186) White female Referred for diagnosis of her cardiac abnormality in 1947, at five and a half years of age.

History Cyanosis was noted at birth and had persisted throughout life A murmur was first heard at three weeks of age Growth and development were slow She sat alone at nine months and walked at two years At three years of age she frequently squatted to rest but soon outgrew the habit

Physical examination Temperature 37°C, pulse 120 per minute, respirations 30 per minute, height 110 cm, weight 15 6 kg, and blood pressure 100/80 mm of mercury

The child was an intelligent, moderately cyanotic, poorly developed girl who suffered from dyspnea at rest There was suffusion of the conjunctivae The lips and buccal mucous membranes were deeply cyanotic The fingers and the toes were cyanotic and clubbed The heart was slightly enlarged The rhythm was regular A systolic murmur of moderate intensity was audible over the precordium, no thrill could be felt The lungs were clear to percussion and auscultation The liver and spleen were not palpable The pulse in the femoral artery was of good quality

Laboratory data Red blood cell count 9 3 million, hemoglobin 23 5 gm, hematocrit 77 per cent, oxygen saturation of the arterial blood 57 per cent

Teleoroentgenogram The heart was slightly enlarged There was fullness of the pulmonary conus and the hilar markings were increased (see Figure XI-4) Upon fluoroscopy faint expansile pulsations were visible in the hilar shadows Delineation of the esophagus with a barium-opaque mixture showed a left aortic arch and no evidence of left auricular enlargement

Electrocardiogram This showed a normal sinus mechanism, sinus tachycardia, high P waves in Lead II, right axis deviation, and evidence of right ventricular hypertrophy (see Figure XI-5)

Clinical impression The clinical findings were characteristic of an Eisenmenger complex in that there was cyanosis, clubbing, and polycythemia the heart was slightly enlarged with X ray evidence of fullness of the pulmonary conus and increased hilar shadows, which upon fluoroscopy showed faint expansile pulsations Nevertheless, the fact that cyanosis dated from birth made us suspect some totally different malformation For this reason special studies were undertaken

Cardiac catheterization revealed an oxygen content of 14 7 volumes per cent in the right auricle, 20 6 volumes per cent in the right ventricle, and 25 0 volumes per cent in the pulmonary artery, whereas the oxygen content in the aorta was only 17 4 volumes

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5 Schultz F B An unusual congenital cardiac anomaly with associated anomalous venous return sudden death American Practitioner and Digest of Treatment 3 616–622 1952 Philadelphia Lippincott

6 Baffes T G A new method for surgical correction of transposition of the aorta and pulmonary artery Surg Gynec & Obst 102 227–233 1956

pulmonary artery not only received blood from the right ventricle but also received blood directly from the left ventricle The pulmonary artery and its branches were greatly dilated The pulmonary orifice measured 5 8 cm in circumference and the main pulmonary artery above the ring measured 6 5 cm The left and the right main branches were approximately 4 0 cm in circumference The pulmonary wall was thicker than normal, its intima, however, was smooth The ductus arteriosus was closed Examination of the myocardium showed that the fibers were hypertrophied but that there were no infarcts and no thrombi The coronary arteries appeared to be normal The bronchial arteries were not enlarged

The lungs were air-containing and showed no evidence of pneumonia or pulmonary infarcts, all the pulmonary vessels were patent Microscopic examination of the lungs revealed occasional thrombi, some of which were in the process of recanalization Many of the small pulmonary arterioles showed diffuse, intimal proliferation which rendered these vessels extremely narrow The lesion appeared to be sufficient to account for the increased resistance in the pulmonary vascular bed In addition, the pulmonary alveoli showed areas of emphysema and areas of atelectasis

The liver showed marked congestion it weighed 1,916 gm The spleen was enlarged and congested, it weighed 140 gm There were many small accessory spleens. The kidneys were normal except for congestion each weighed 80 gm The cortex and medulla were well defined The pelvis and ureters were normal There was diffuse hemorrhage in the thymus

Final anatomical diagnosis Transposition of the aorta Dilatation and slight displacement of the pulmonary artery Ventricular septal defect Foramen ovale covered by a valve, but not completely sealed Dilatation and hypertrophy of the right ventricle Extreme thickening and intimal proliferation of the pulmonary arterioles and small pulmonary arteries Occasional recanalization of thrombi in the pulmonary arterioles Splenomegaly Patchy emphysema and atelectasis Acute congestion of the lungs and viscera Diffuse hemorrhage in the thymus

Comment This was the first malformation of its type which the author had studied Diagnosis was correct in that the aorta arose from the right ventricle and the pulmonary artery occupied its normal position The fact that the pulmonary artery could be levoposed was not known to the author but the possibility should have been considered, because the oxygen content of the blood in the pulmonary artery was higher than that in the right ventricle and was also significantly higher than that in the aorta

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cle pumped out 30 cc of blood, part of the blood would go to the lungs and part to the lower extremities Let us suppose that 20 cc go to the lungs and 10 cc. to the lower extremities The 20 cc which are directed to the lungs will be returned to the left auricle and this volume of blood will be pumped *out from the left* ventricle into the aorta All the blood in the aorta goes to the *head and the upper* extremities This blood will be returned to the right auricle by the superior vena cava In addition, the 10 cc of blood which were pumped from the right ventricle through the ductus arteriosus to the lower extremities will be returned to the right auricle by the inferior vena cava Therefore the right auricle will receive a total of 30 cc This amount will pass into the right ventricle Then the cycle starts again The course of the circulation is shown in Diagram XII-1

PHYSIOLOGY OF THE MALFORMATION

In this malformation the right ventricle not only pumps the blood to the lungs but also to the trunk and the lower extremities Consequently the pressure in the pulmonary artery is the same as that in the lower extremities It follows that the pressure in the lungs is unduly high Therefore, although the lungs receive venous blood from the right ventricle there is marked pulmonary hypertension Furthermore inasmuch as the right ventricle pumps all the blood which goes to the lungs and also all the blood which goes to the trunk and the lower extremities the right side of the heart carries a greater load than the left

CLINICAL FINDINGS

The difference in cyanosis between the upper and the lower extremities is the feature of diagnostic importance The head and the upper extremities receive oxygenated blood and therefore are not cyanotic, whereas the lower extremities receive venous blood which is ordinarily destined to go to the lungs for oxygenation and therefore are cyanotic The line of demarcation of the cyanosis lies at the brim of the pelvis, as it does in cases of complete transposition of the great vessels and not high up on the shoulders, as in cases of *pulmonary hypertension* with persistent patency of the ductus arteriosus In the malformation under discussion the left subclavian artery usually arises from the ascending aorta and carries fully oxygenated blood Inasmuch as there is a complete interruption of the isthmus of the aorta, no blood from the pulmonary artery can enter the subclavian artery, hence the cyanosis appears at a low level If, *however, the left* subclavian artery arises from the descending aorta, the line of demarcation of the cyanosis will be high up across the shoulders (see Chapter XVIII, Section B)

mation, the blood from the left ventricle is directed by way of the aorta to the head and the upper extremities and the blood from the right ventricle is pumped into the pulmonary artery and through the ductus arteriosus into the descending aorta The condition is readily compatible with fetal life and places no strain on the fetal circulation (see Figure XII–2)

After birth, however, the situation is quite different The blood from the left ventricle is pumped out by way of the aorta to the head and the upper extremities and is returned by the superior vena cava to the right auricle and to the right ventricle, thence it is pumped into the pulmonary artery Part of the blood from the pulmonary artery passes into the lungs and part through the ductus arteriosus to the descending aorta and the lower extremities The blood from the lungs is returned to the left auricle and the left ventricle, the blood from the lower extremities is returned by way of the inferior vena cava to the right auricle

Consequently, with each cardiac cycle, the right side of the heart receives a greater volume of blood than does the left side For example, if the right ventri

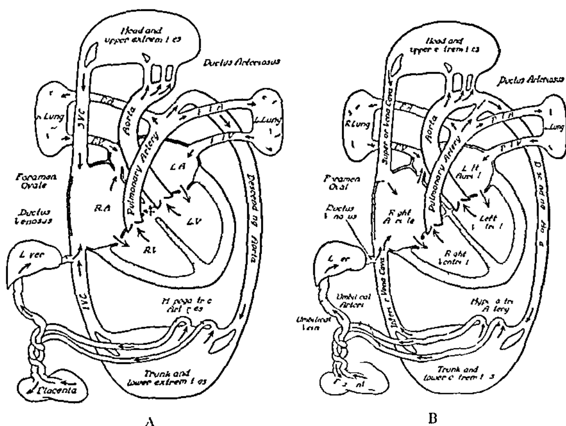


FIGURE XII–2 Fetal circulation (A) Complete interruption of the isthmus of the aorta and a persistent ostium atrioventriculare commune and (B) normal heart

cle pumped out 30 cc of blood, part of the blood would go to the lungs and part to the lower extremities Let us suppose that 20 cc. go to the lungs and 10 cc to the lower extremities The 20 cc which are directed to the lungs will be returned to the left auricle and this volume of blood will be pumped out from the left ventricle into the aorta All the blood in the aorta goes to the head and the upper extremities This blood will be returned to the right auricle by the superior vena cava In addition, the 10 cc of blood which were pumped from the right ventricle through the ductus arteriosus to the lower extremities will be returned to the right auricle by the inferior vena cava Therefore the right auricle will receive a total of 30 cc. This amount will pass into the right ventricle Then the cycle starts again The course of the circulation is shown in Diagram XII-1

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CLINICAL FINDINGS

The difference in cyanosis between the upper and the lower extremities is the feature of diagnostic importance The head and the upper extremities receive oxygenated blood and therefore are not cyanotic, whereas the lower extremities receive venous blood which is ordinarily destined to go to the lungs for oxygenation and therefore are cyanotic The line of demarcation of the cyanosis lies at the brim of the pelvis, as it does in cases of complete transposition of the great vessels, and not high up on the shoulders, as in cases of pulmonary hypertension with persistent patency of the ductus arteriosus In the malformation under discussion the left subclavian artery usually arises from the ascending aorta and carries fully oxygenated blood Inasmuch as there is a complete interruption of the isthmus of the aorta no blood from the pulmonary artery can enter the subclavian artery hence the cyanosis appears at a low level If however the left subclavian artery arises from the descending aorta, the line of demarcation of the cyanosis will be high up across the shoulders (see Chapter XVIII, Section B)

DIAGRAM XII–1

Ductus Arteriosus

R Lung

L Lung

S.V.C.

P.A.

Aorta

pul A.

PV

Pul V

Pulmonary Artery

L A

R.A.

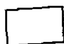
LV

RV

Descending Aorta

I.V.C.

Trunk and lower extremities



Arterial blood (fully saturated)



Small admixture of venous blood
No visible cyanosis



Venous and arterial blood
Cyanosis visible



Venous blood

Diagram XII–1

Absence of the aortic arch (complete interruption of the isthmus of the aorta) and a persistent ostium atrioventriculare commune The descending aorta is continuous with the pulmonary artery through the ductus arteriosus

The essential feature of this malformation is the complete interruption of the isthmus of the aorta The descending aorta is continuous with the pulmonary artery through the ductus arteriosus It follows that the pressure in the pulmonary artery is the same as that in the descending aorta Consequently there is marked pulmonary hypertension

It may be that a persistent ostium atrioventriculare commune or a ventricular septal defect is also an integral part of this malformation The significant changes in circulation however, depend primarily upon the complete interruption of the isthmus of the aorta

The blood from the right auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs and through the ductus arteriosus into the descending aorta The blood from the lungs is returned to the left auricle and thence to the left ventricle and is pumped out through the aorta to the head and the upper extremities The blood from the head and the upper extremities is returned by the superior vena cava to the right auricle and the blood from the trunk and the lower extremities is returned by the inferior vena cava to the right auricle Therefore, with each cardiac cycle the right auricle receives all the blood which was pumped out from the left ventricle to the head and the upper extremities and also that part of the blood from the right ventricle which was pumped from the pulmonary artery through the ductus arteriosus to the trunk and the lower extremities The left auricle receives only that part of the blood from the right ventricle which was directed through the pulmonary artery to the lungs Therefore the pressure in the right auricle is greater than the pressure in the left auricle Some of the blood flows from the right auricle into the left auricle and some of the blood flows from the right ventricle into the aorta If the pressure on the left side of the heart rises the direction of the shunt will be reversed The circulation of the blood within the heart is similar to that *in a persistent ostium* atrioventriculare commune except that the pressure tends to remain higher in the right side of the heart as more blood is always returned to the right auricle than to the left auricle

Clinical diagnosis The most important diagnostic clue is the difference in cyanosis between the upper and the lower extremities The upper extremities receive oxygenated blood from the left side of the heart whereas the lower extremities receive venous blood from the right side of the heart because the descending aorta is continuous with the pulmonary artery Therefore the lower extremities will be cyanotic whereas the upper extremities will show no cyanosis The occurrence of a septal defect may or may not be an integral part of the malformation A murmur and a thrill if present are strongly suggestive of some additional defect

If the malformation is combined with a persistent ostium atrioventriculare commune, the difference in the intensity of the cyanosis between the upper and the lower extremities may be slight and will call for careful observation It is always best appreciated by placing the patient's hand beside his foot If the face and hands are of normal hue, the cyanosis of the feet may be overlooked Furthermore, if noted, it is likely to be attributed to cold or to a sluggish circulation The fact that the feet remain cyanotic when warm offers the clue to the correct diagnosis

The pulse is of good quality and of equal strength in the arm and the leg In contrast to coarctation of the aorta, this anomaly causes no obstruction to the flow of blood to the lower extremities The pulse in the lower extremities is normal

CARDIAC FINDINGS

The heart is slightly to moderately enlarged The enlargement affects the right side, it is due to the increased volume of blood which is returned to the right auricle and the right ventricle If the enlargement of the right ventricle is sufficiently great so that the right ventricle presses against the anterior chest wall, there will be left sided chest deformity

Murmurs may or may not be present

X RAY AND FLUOROSCOPIC FINDINGS

The heart is enlarged to the right and to the left Owing to the normal position of the pulmonary artery, the shadow at the base of the heart to the left of the sternum will be of normal contour Indeed, the pulmonary conus may be abnormally full

In the left anterior oblique position the enlargement of the right ventricle will be demonstrated by the projection of the anterior border of the cardiac silhouette toward the anterior chest wall The left ventricle will be seen to be small, it will require but little rotation of the patient for the left ventricle to clear the spinal column

ELECTROCARDIOGRAPHIC FINDINGS

The standard leads show a right axis deviation and the unipolar precordial leads show evidence of right ventricular hypertrophy

DIAGNOSIS

The diagnosis is based upon the distribution of the cyanosis The head and

the upper extremities are of normal color, whereas the lower extremities are cyanotic because the descending aorta receives its blood from the pulmonary artery through the ductus arteriosus

DIFFERENTIAL DIAGNOSIS

The common error is the failure to appreciate the significance of the cyanosis of the feet If the head and the upper extremities are of good color, ordinarily little consideration is given to the circulation to the lower extremities Consequently the possibility of this abnormality is missed Even if cyanosis is noted, its significance is often not appreciated

TREATMENT

This abnormality could be corrected by the insertion of a graft between the ascending aorta and the descending aorta combined with the division of the ductus arteriosus

PROGNOSIS

The prognosis is usually poor If the condition should occur as an isolated anomaly successful surgery would restore the circulation to normal Usually, however the prognosis is determined by the associated anomaly, which may or may not be amenable to surgery

SUMMARY

The characteristic feature of a complete interruption of the isthmus of the aorta, in which the descending aorta becomes continuous with the pulmonary artery through the ductus arteriosus is that the cyanosis is confined to the lower extremities The pulse in both the upper and the lower extremities is strong The increased volume of blood which is pumped through the right auricle and the right ventricle produces enlargement of the right side of the heart, this in turn, may lead to left sided chest deformity

This malformation, if not associated with other anomalies, could be corrected by surgery

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CHAPTER XIII

DEFECTIVE DEVELOPMENT OF THE LEFT SIDE OF THE HEART

ATRESIA or marked hypoplasia of the aortic orifice is always associated with hypoplasia of the ascending aorta and also with defective development of the left ventricle In addition there may or may not be atresia of the mitral valve These conditions are discussed in Section A Section B presents an entirely different defect of the left ventricle, namely, a diverticulum of the left ventricle

A *Aortic Atresia or Marked Hypoplasia of the Aortic Orifice Combined with Hypoplasia of the Ascending Aorta*

This malformation is in a sense the counterpart of tricuspid atresia and a non functioning right ventricle Atresia or marked hypoplasia of the tricuspid valve is always associated with defective development of the right ventricle, the atresia of the pulmonary orifice is secondary to the failure of the right ventricle to function The aortic atresia and the marked hypoplasia of the ascending aorta are, however, the primary features of the malformation under discussion and the underdevelopment of the left ventricle and the mitral atresia are usually secondary to the abnormality of the aorta Thus in the former the primary abnormality concerns the tricuspid valve, which directs the blood to the right ventricle, whereas in the latter malformation the primary difficulty is the egress of blood from the left ventricle

NATURE OF THE MALFORMATION

The essential feature of this malformation is the atresia of the aortic valves or the marked hypoplasia of the aortic orifice combined with hypoplasia of the aorta up to the point of entrance of the ductus arteriosus Whether the aortic orifice is atretic or markedly hypoplastic, the underdevelopment of the aorta is always associated with an imperfect development of the left ventricle, either the left ventricle is absent or it is a thin walled, poorly developed chamber

The absence or marked hypoplasia of the left ventricle in turn affects the egress of blood from the left auricle When there is no left ventricle, the mitral

valve is always atretic and there is also a gross defect in the auricular septum When the aorta is markedly hypoplastic and the left ventricle is a relatively thin walled chamber, it is also common to find a small defect in the auricular septum In either case the main flow of blood to the systemic circulation is from the left auricle to the right auricle and thence to the right ventricle, from there the blood is pumped into the pulmonary artery and through the ductus arteriosus to the aorta The aorta, beyond the entrance of the ductus arteriosus, is usually of normal caliber The aorta, proximal to the entrance of the ductus arteriosus, always persists as a diminutive tube, and it is from the base of the aorta that the coronary arteries originate In cases of aortic atresia, the coronary arteries, and indeed the head and the upper extremities, receive their nutrition from the back flow of blood fom the ductus arteriosus to the base of the aorta When the aorta persists as a diminutive tube, the coronary arteries receive their supply of blood from the diminutive aorta in the normal manner Nevertheless, this malformation always causes real difficulty in the expulsion of blood from the left ventricle the main supply of blood to the systemic circulation is from the pulmonary artery through the ductus arteriosus Figure XIII-1 illustrates a case of aortic atresia mitral atresia and absence of the left ventricle Figure XIII-2 illustrates a case of marked hypoplasia of the aorta with a relatively thin walled left ventricle the aorta beyond the entrance of the ductus arteriosus is of normal caliber

Atresia of the aortic orifice and marked hypoplasia of the ascending aorta are always extremely serious Fortunately, although anatomically familiar, these conditions are clinically rare

The rarity of this malformation is in all probability due to the functional importance of the left ventricle during fetal life An embryo in which the left ventricle and aorta fail to develop can remain viable only if the function of the left ventricle is taken over by the right It is probable that in many instances the fetus dies at an early age In most instances the malformation is compatible with extra uterine life only for a short time

The condition is obviously more likely to be compatible with life when a defective left ventricle is combined with hypoplasia of the ascending aorta than when the aortic orifice is atretic Indeed, when the left ventricle is defective, the condition merges into that of pulmonary hypertension with persistent patency of the ductus arteriosus secondary to a severe left sided cardiac lesion In the older age group the signs and symptoms are primarily due to the pulmonary hypertension and the reversal of the flow of blood through the ductus arteriosus (see Chapter XVIII, Section B)

CHAPTER XIII

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ATRESIA or marked hypoplasia of the aortic orifice is always associated with hypoplasia of the ascending aorta and also with defective development of the left ventricle In addition there may or may not be atresia of the mitral valve These conditions are discussed in Section A Section B presents an entirely different defect of the left ventricle, namely, a diverticulum of the left ventricle

A *Aortic Atresia or Marked Hypoplasia of the Aortic Orifice Combined with Hypoplasia of the Ascending Aorta*

This malformation is in a sense the counterpart of tricuspid atresia and a non functioning right ventricle Atresia or marked hypoplasia of the tricuspid valve is always associated with defective development of the right ventricle, the atresia of the pulmonary orifice is secondary to the failure of the right ventricle to function The aortic atresia and the marked hypoplasia of the ascending aorta are, however, the primary features of the malformation under discussion and the underdevelopment of the left ventricle and the mitral atresia are usually secondary to the abnormality of the aorta Thus in the former the primary abnormality concerns the tricuspid valve, which directs the blood to the right ventricle, whereas in the latter malformation the primary difficulty is the egress of blood from the left ventricle

NATURE OF THE MALFORMATION

The essential feature of this malformation is the atresia of the aortic valves or the marked hypoplasia of the aortic orifice combined with hypoplasia of the aorta up to the point of entrance of the ductus arteriosus Whether the aortic orifice is atretic or markedly hypoplastic, the underdevelopment of the aorta is always associated with an imperfect development of the left ventricle, either the left ventricle is absent or it is a thin walled, poorly developed chamber

The absence or marked hypoplasia of the left ventricle in turn affects the egress of blood from the left auricle When there is no left ventricle, the mitral

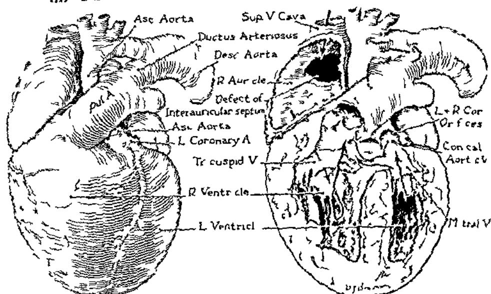


FIGURE XIII-2 Extreme aortic stenosis and hypoplasia of the ascending aorta with underdevelopment of the left ventricle, a high ventricular septal defect, and a gross defect in the auricular septum

COURSE OF THE CIRCULATION

During fetal life a considerable portion of the blood which enters the right auricle normally flows through the foramen ovale to the left auricle and thence to the left ventricle. This blood is pumped through the aorta to the body of the fetus. In cases of aortic atresia and a non functioning left ventricle, if the fetus is to remain alive, the work of the left ventricle must be taken over by the right ventricle. Under such circumstances all the blood from the right auricle flows into the right ventricle and is pumped out into the pulmonary artery. The major part of the blood which enters the pulmonary artery passes in the normal manner by way of the ductus arteriosus to the body of the fetus. The volume of blood which is pumped through the ductus arteriosus is, however, far greater than normal. Virtually the entire work of the heart is performed by the right ventricle. For this reason during intra uterine life the right ventricle undergoes great hypertrophy. Indeed this is one of the few malformations in which the infant is born with enormous right sided cardiac enlargement. Figure XIII-3 shows the fetal circulation in aortic atresia and Figure XIII-4 shows the fetal circulation in defective development of the left ventricle and marked hypoplasia of the ascending aorta.

At birth with the expansion of the lungs there is an abrupt fall in the pul

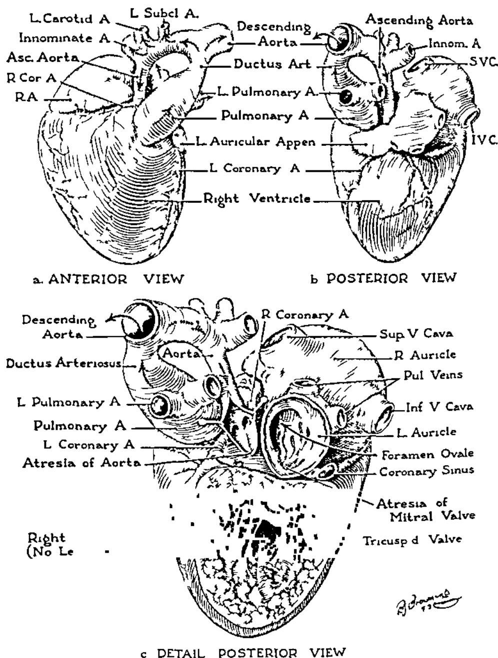


FIGURE XIII-1 Aortic atresia and marked hypoplasia of the ascending aorta, mitral atresia and absence of the left ventricle

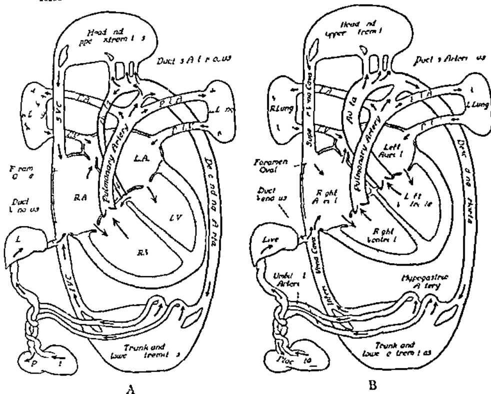


FIGURE XIII–4 Fetal circulation (A) Extreme aortic stenosis marked hypoplasia of the ascending aorta an underdeveloped left ventricle, and auricular and ventricular septal defects and (B) normal heart

non functioning, the blood cannot leave the left auricle in the normal manner All the blood from the left auricle must escape through a defect in the auricular septum to the right auricle

Thus the oxygenated blood which is returned from the lungs to the left auricle encounters difficulty in reaching the systemic circulation In the majority of other malformations cyanosis is due to the shunting of venous blood into the *systemic circulation and thereby the oxygenated blood is diluted with venous* blood In the malformation under discussion the body is primarily supplied with venous blood Furthermore, it is difficult for the oxygenated blood to reach the systemic circulation The course of the circulation is shown in Diagram XIII–1 Diagram XIII–2 shows the course of the circulation in a case of marked hypoplasia of the ascending aorta and defective development of the left ventricle

PHYSIOLOGY OF THE MALFORMATION

The right ventricle maintains the systemic circulation by pumping the blood

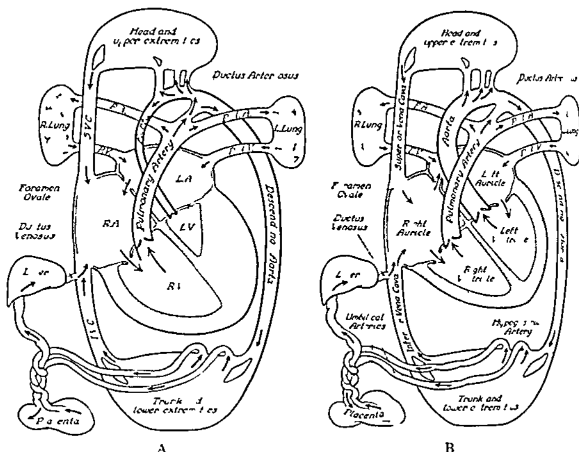


FIGURE VIII–3 Fetal circulation (A) Aortic atresia, mitral atresia, a non functioning left ventricle, and an auricular septal defect and (B) normal heart

monary pressure and consequently the blood in the pulmonary artery is directed to the lungs The sudden diversion of the blood to the lungs causes a cessation of the flow of blood through the ductus arteriosus to the aorta The consequence is that the systemic circulation, which has received its blood supply through this channel, is momentarily deprived of blood Not until the pressure in the pulmonary circulation becomes greater than that in the systemic circulation will the blood again flow through the ductus arteriosus to the aorta Then, and then only, will the systemic circulation be reestablished Moreover, upon the restoration of the circulation, the blood flows from the pulmonary artery to the aorta, the body is supplied with venous blood which is ordinarily destined to go to the lungs for oxygenation The blood which goes to the systemic circulation is returned by way of the superior and inferior venae cavae to the right auricle and thence it again passes to the systemic circulation The blood from the lungs is returned by way of the pulmonary veins to the left auricle If the left ventricle is absent or

DIAGRAM XIII-1

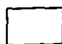
Aortic atresia and marked hypoplasia of the ascending aorta combined with a patent ductus arteriosus, mitral atresia, a non functioning left ventricle and an auricular septal defect

The essential feature in this malformation is the atresia of the aorta at its base. The left ventricle does not function The left ventricle is either entirely absent or represented by a diminutive chamber filled with a blood clot. The mitral valve is completely atretic or markedly hypoplastic In addition there is always a defect in the auricular septum and the ductus arteriosus is patent. The blood supply to the systemic circulation is from the pulmonary artery through the ductus arteriosus The coronary arteries arise from the base of the aorta in the normal fashion but receive no blood from the left ventricle The blood supply to the myocardium is from the back flow of blood from the transverse arch of the aorta

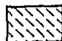
The blood from the right auricle passes into the right ventricle and is pumped out by way of the pulmonary artery to the lungs and through the ductus arteriosus to the systemic circulation The blood from the lungs is returned to the left auricle. Inasmuch as there is an atresia of the mitral valve and the left ventricle does not function, the only way for the blood to escape from the left auricle is through the defect in the auricular septum to the right auricle Thence it passes to the right ventricle and is pumped out into the pulmonary artery Inasmuch as no blood is pumped from the left ventricle into the aorta, the pressure in the systemic circulation is low and blood flows from the pulmonary artery through the ductus arteriosus into the aorta This is the only way for the blood to reach the systemic circulation The coronary arteries arise from the base of the aorta and are fed by the blood which flows back from the ductus arteriosus to the ascending aorta and down to the base of the aorta The greater the volume of blood which reaches the lungs for oxygenation the greater is the deprivation of the systemic circulation At best the systemic circulation is supplied with a *mixture of venous and arterial blood* from the right ventricle. The circulation is very inefficient. The blood from the systemic circulation is returned in the normal manner by the superior and inferior venae cavae to the right auricle and to the right ventricle. The right ventricle pumps the blood not only to the lungs but also to the systemic circulation Hence there is always severe pulmonary hypertension Great hypertrophy of the right ventricle results

The blood in both the right auricle and the right ventricle is a mixture of the venous blood returned from the systemic circulation and of the oxygenated blood returned from the lungs With the complete expansion of the lungs the blood in the pulmonary artery is directed to the lungs and, although a greater volume of blood is returned to the left auricle, the blood is directed away from the systemic circulation until the pressure in the pulmonary circulation exceeds that in the systemic circulation

DIAGRAM XIII-1


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

from the pulmonary artery through the *ductus arteriosus* to the aorta Consequently the pressure in the pulmonary artery is the same as that in the systemic circulation Hence there is always *pulmonary hypertension* The only blood which the body ever receives is the blood which is pumped from the pulmonary artery through the *ductus arteriosus* to the aorta It is, however, important to remember that the blood in the pulmonary artery contains the oxygenated blood which has been returned by the pulmonary veins to the left auricle and shunted through the auricular defect to the right auricle, where it mixes with the venous blood returned by the superior and inferior venae cavae This mixture of arterial and venous blood flows into the right ventricle and is pumped out into the pulmonary artery and through the ductus arteriosus to the systemic circulation In the absence of an auricular defect, great difficulty is encountered in the expulsion of blood from the left auricle The increased pressure in the left auricle increases the resistance in the lungs and increases still further the work required of the right ventricle

CLINICAL FINDINGS

Cyanosis is always intense The baby is blue at birth In contrast to almost all other malformations the first breaths of life do nothing to relieve the cyanosis Indeed the establishment of respiration directs the blood to the lungs and away from the systemic circulation The only blood which the systemic circulation ever receives is the blood from the pulmonary artery which is ordinarily destined to go to the lungs for oxygenation Moreover the oxygenated blood which is returned from the lungs to the left auricle encounters difficulty in reaching the body Inasmuch as the aortic orifice is atretic and the entire systemic circulation is maintained through the ductus arteriosus, cyanosis is of uniform distribution

Dyspnea is severe

The pulse is so weak that it is frequently impalpable When palpable, it is of equal strength in the arm and the leg

The blood pressure is low

Engorgement of the liver and *edema* occur early

CARDIAC FINDINGS

The heart is enormously enlarged to the right and to the left Indeed, hypoplasia of the ascending aorta combined with defective development of the left ventricle is virtually the only malformation which causes the heart to be greatly enlarged at birth Figure XIII-5 shows the size of the heart in an infant twenty

Cyanosis is intense The condition is rarely compatible with life for more than a few days

Clinical diagnosis is based upon the early occurrence of intense cyanosis, weak pulses, great cardiac enlargement with forceful heart sounds, and right sided cardiac failure X ray or fluoroscopic examination shows that the enlargement is primarily due to the enlargement of the right ventricle In the anterior posterior position there is great enlargement of the pulmonary conus and in the left anterior-oblique position the right ventricle may be seen to extend to the anterior chest wall The condition of the infant remains critical throughout its brief life Death usually occurs within the first weeks of life

DIAGRAM XIII–2

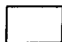
Extreme aortic stenosis and marked hypoplasia of the ascending aorta combined with an underdeveloped left ventricle a patent ductus arteriosus and auricular and ventricular septal defects

This malformation closely resembles that of aortic atresia The aorta, however, remains as a diminutive tube which may arise from the thin walled left ventricle or may over ride both ventricles In any event the main circulation to the body is by way of the right side of the heart, the systemic circulation receives its blood from the pulmonary artery through the ductus arteriosus Consequently the systolic pressure in the right ventricle must be equal to the systemic pressure Hence there is a marked pulmonary hypertension

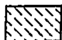
The blood from the right auricle flows into the right ventricle. By far the greater part of the blood is pumped out into the pulmonary artery, thence the blood flows to the lungs and through the ductus arteriosus to the systemic circulation The blood from the lungs is returned in the normal fashion by the pulmonary veins to the left auricle A small portion of the blood flows from the left auricle into the left ventricle and is pumped out through the stenosed aortic orifice into the diminutive aorta The greater part of the blood from the left auricle flows through the defect in the auricular septum to the right auricle where it mixes with the blood which is returned by the superior and inferior venae cavae to the right auricle Virtually all the blood from the right auricle flows into the right ventricle and is pumped out into the pulmonary artery When the diminutive aorta over rides the ventricular septum, a small volume of blood from the right ventricle can be pumped directly into the aorta, nevertheless, the aorta is so small that the systemic circulation receives but a minimal amount of blood from the diminutive aorta The coronary arteries, however, arise from the base of the aorta in the normal fashion and receive their blood from the small volume of blood which is pumped into the diminutive ascending aorta

Clinical diagnosis The clinical findings are essentially the same as those of aortic atresia The infant is born with great right sided cardiac enlargement the x ray shows marked fullness of the pulmonary conus, cyanosis is intense. Cardiac failure occurs early the condition is compatible with life for only a few days

Diagram XIII–2


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

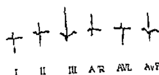
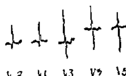


FIGURE XIII–6 Defective development of the left ventricle and hypoplasia of the ascending aorta (same patient as in Figure XIII–5) Infant



ular septum is small, there may be enlargement of the left auricle The duration of life is usually too short to permit great enlargement of the left auricle The baby is often too feeble to swallow barium and consequently no evidence can be obtained concerning the size of the left auricle

ELECTROCARDIOGRAPHIC FINDINGS

The standard leads show a right axis deviation and the unipolar leads show evidence of marked right ventricular hypertrophy The T waves may be of low amplitude (see Figure XIII–6)

SPECIAL TESTS

The condition of the infant is generally too critical to permit more than a brief examination

Cardiac catheterization would reveal a left to-right shunt at the auricular level and systemic pressure in both the right ventricle and the pulmonary artery It would probably be possible to catheterize the descending aorta through the ductus arteriosus

Angiocardiography would reveal simultaneous opacification of the descending aorta and the pulmonary artery

DIAGNOSIS

The diagnosis is based upon the findings of intense cyanosis, enormous right sided cardiac enlargement which dates from birth, the forcefulness of the heart sounds and the weakness of the pulses, and early signs of right sided cardiac failure



FIGURE VIII-5 Defective development of the left ventricle and hypoplasia of the ascending aorta (same patient as in Figure VIII-6) Infant twenty four hours of age

four hours old In this instance the enormous size of the cardiac shadow, detected when the infant was twelve hours of age, had raised the question of a mediastinal tumor *The heart sounds* are forceful *Murmurs* may or may not be present *Cardiac failure* occurs early and within the first two or three days of life the liver becomes engorged and extends to the umbilicus

X RAY AND FLUOROSCOPIC FINDINGS

Fluoroscopic examination shows great enlargement of the right auricle, of the right ventricle, and of the pulmonary conus In addition, the superior vena cava is often dilated, consequently the shadow at the base of the heart is abnormally wide Upon rotation of the infant s head to the left, the pulmonary artery can be visualized in its normal position The lungs remain relatively clear because the pulmonary vascular bed opens up slowly The heart may be so enormous that only in the left anterior oblique position is it possible to ascertain that the increased size of the heart is primarily due to great enlargement of the right ventricle In this position the anterior border of the cardiac silhouette extends to the anterior chest wall Examination in the right anterior-oblique position reveals little The cardiac shadow appears to fill the entire chest

The size of the left auricle depends upon the size of the auricular defect If there is mitral atresia in addition to the aortic atresia, and the defect in the auric

and a foramen ovale guarded by a valve may cause early signs of distress At birth, however, the heart is normal in size, the enlargement is barely detectable at two days of age

Pulmonary hypertension with reversal of the flow of blood through the ductus arteriosus, when caused by a severe left sided cardiac lesion, may be due to defective development of the left ventricle and marked hypoplasia of the ascending aorta If the condition is not extreme and the left ventricle supplies the blood to the head and the upper extremities, the clinical syndrome is that of pulmonary hypertension and a reversed ductus, in which the trunk and the lower extremities are more cyanotic than the head and the upper extremities (see Chapter XVIII, Section B) When there is atresia of the aortic orifice, the entire circulation is maintained by the right ventricle The work of the right ventricle is greatly increased and the heart is greatly enlarged, cyanosis is intense and of uniform distribution

Truncus arteriosus may be confused with aortic atresia, especially when the circulation to the lungs is by way of the bronchial arteries Under such circumstances cyanosis appears early and is associated with great cardiac enlargement The contour of the heart, however, is different In a truncus arteriosus, the shadow at the base of the heart to the left of the sternum is concave, whereas in aortic atresia the pulmonary conus is abnormally prominent Furthermore, in the right anterior-oblique position the sharp angulation of the right ventricle from the aorta to the anterior chest wall is seen only in a truncus arteriosus (see Chapter XIV) the exaggeration of the pulmonary conus in an aortic atresia causes a diffuse bulge along the anterior margin of the cardiac silhouette

TREATMENT

There is no effective treatment An aortic atresia and a marked hypoplasia of the ascending aorta, combined with a poorly developed left ventricle, constitute such a severe abnormality that surgical correction is impossible

PROGNOSIS

The prognosis is hopeless The condition of the infant remains precarious throughout its brief life Cardiac failure occurs early The malformation is usually compatible with life for only a few days

SUMMARY

The outstanding features of aortic atresia are intense cyanosis, great cardiac enlargement and the early onset of cardiac failure The heart sounds are force

The fluoroscopic findings give the clue to the diagnosis The tremendous size of the right ventricle indicates that the right, not the left ventricle, is the chamber of functional importance in the maintenance of the systemic circulation The enlargement of the pulmonary conus shows that the pulmonary artery is given off the right ventricle in the normal fashion These two findings together indicate that the blood from the right ventricle is pumped through the pulmonary artery to the systemic circulation It follows that the ductus arteriosus must be widely patent and of functional importance in the direction of the blood from the pulmonary artery to the aorta In order for the blood to flow from the pulmonary artery through the ductus arteriosus to the aorta, the pressure in the systemic circulation must be lower than that in the pulmonary circulation Therefore the flow of blood from the left ventricle into the aorta must be meager From these facts it is deduced that the primary malformation is an aortic atresia or marked hypoplasia of the aortic orifice combined with hypoplasia of the ascending aorta

Inasmuch as this malformation is always associated with underdevelopment of the left ventricle, the blood cannot leave the left auricle in the normal manner Consequently there must be some defect in the auricular septum The clinical manifestations of the auricular septal defect vary according to its size and may be difficult to determine If the defect is large, the thin walled, readily distensible right auricle bears the brunt of the increased auricular pressure This, too, can be detected upon fluoroscopy by the distention of the superior vena cava If the defect is small, the left auricle becomes enlarged and causes backward displacement of the esophagus

DIFFERENTIAL DIAGNOSIS

Aortic atresia calls for differentiation from other anomalies which place great strain on the right side of the heart and from other conditions which are fatal in early infancy

Total anomaly of the pulmonary venous return in the terminal stage may be confused with aortic atresia, because in both instances in early infancy the patient may be intensely cyanotic The history is usually of aid, as a baby with a total anomaly of the venous return is seldom intensely cyanotic from birth except when all the pulmonary veins drain into the hepatic vein, under such circumstances the heart is phenomenally small

"Pure" pulmonary stenosis may cause cyanosis and cardiac failure in the neonatal period Furthermore, at this age the heart is but slightly enlarged The infant with "pure" pulmonary stenosis usually responds promptly to digitalis

Complete transposition of the great vessels with a closed ventricular septum

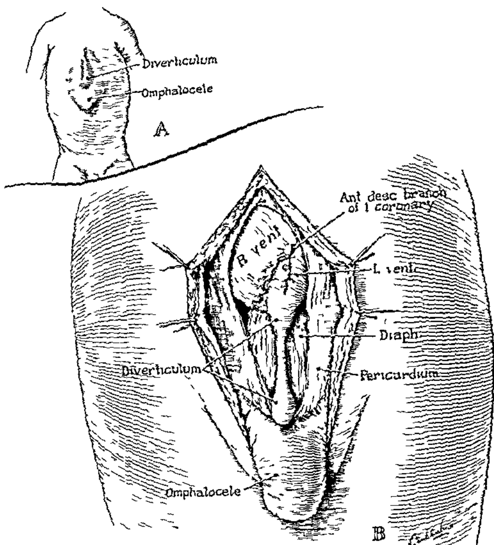


FIGURE XIII-7 Diverticulum of the left ventricle, Case XIII-1

heart with increased pulmonary blood flow The pulsating vessel in the abdominal wall was thought to be a diverticulum of the left ventricle but the possibility of an aberrant vessel arising from the descending aorta was also considered

Special tests Angiocardiography was performed in the usual manner The right side of the heart filled normally Then the left side of the heart and the aorta were visualized There was slight opacification below the diaphragm at the time the dye entered the left ventricle but *it was not clearly delineated The descending aorta* was clearly delineated and it was obvious that no abnormal vessel arose therefrom

ful Murmurs may or may not be present The pulse is weak but, if palpable, is of equal intensity in the arm and the leg Fluoroscopic examination shows that the enlargement is primarily right sided and that the pulmonary conus is abnormally full The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy

The condition calls for differentiation from other malformations which cause cardiac failure in the neonatal period and are compatible with life for only a short time A total anomaly of the venous return is sometimes confused with this malformation Pulmonary stenosis with an intact ventricular septum and complete transposition of the great vessels with a closed ventricular septum may cause cardiac failure in the neonatal period Congenital mitral atresia may closely simulate this malformation There is as yet no treatment The prognosis is hopeless Death usually occurs within the first few days

B *Diverticulum of the Left Ventricle*

A diverticulum which extends from the apex of the left ventricle through the diaphragm into the abdomen is a rare abnormality, but several cases have been reported[1,3] It is usually associated with an omphalocele and is frequently associated with tricuspid atresia The diagnosis is made by the pulsating mass in the upper abdomen The following case report illustrates the condition

Illustrative Case

Case VIII-1 Baby K (Harriet Lane Home, No A-96602) White male First seen in the Cardiac Clinic in September, 1952, at two months of age because of a heart murmur

Physical examination The patient was well nourished and well-developed and had good color The head and neck showed no abnormalities Examination of the abdomen revealed a large omphalocele (see Figure VIII-7) and, within its upper portion, a vessel which pulsated vigorously and extended up to the diaphragm The vessel felt like a thick walled artery about 3 mm in diameter It was possible to slide one s fingers beneath the distal end of this vessel It was also possible to constrict the vessel in its mid portion, when this was done both the proximal and the distal segments pulsated vigorously

Examination of the heart revealed slight cardiac enlargement The heart sounds were of good quality The pulmonic second sound was accentuated and there was a harsh precordial systolic murmur

Fluoroscopy revealed slight cardiac enlargement and increased hilar shadows

Clinical impression The baby obviously had a congenital malformation of the

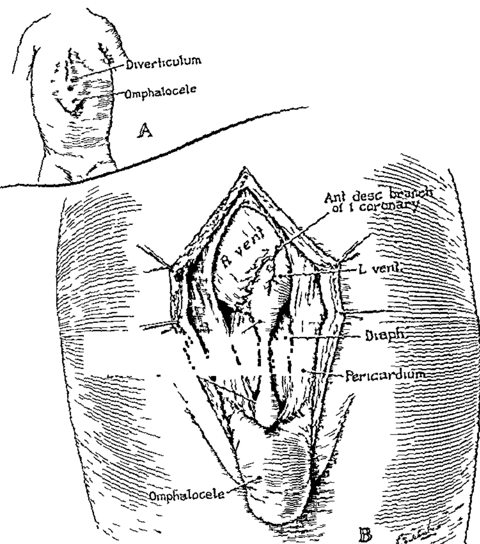


FIGURE XIII-7 Diverticulum of the left ventricle Case XIII-1

heart with increased pulmonary blood flow The pulsating vessel in the abdominal wall was thought to be a diverticulum of the left ventricle but the possibility of an aberrant vessel arising from the descending aorta was also considered

Special tests Angiocardiography was performed in the usual manner The right side of the heart filled normally Then the left side of the heart and the aorta were visualized There was slight opacification below the diaphragm at the time the dye entered the left ventricle but it was not clearly delineated The descending aorta was clearly delineated and it was obvious that no abnormal vessel arose therefrom

Clinical diagnosis Congenital malformation of the heart and a diverticulum of the left ventricle

Disposition Although in 1952 surgical correction of the cardiac malformation was not possible, it was hoped that removal of the diverticulum would strengthen the left ventricle Therefore surgery was recommended

Treatment Surgical correction was attempted but, as soon as the diverticulum was dissected and a clamp was placed on it, the heart stopped and could not be restarted

Autopsy No 23910 (limited, performed by Dr Morgan Berthrong) The external examination of the infant revealed that the umbilicus was absent and that there was a protrusion of the anterior abdominal wall which formed a hernial pouch 1 5 cm below the tip of the diaphragm This mass measured 6 x 9 cm There was a surgical incision which extended down the mid portion of the mass When the surgical incision was opened, it was apparent that a portion of the abdominal mass was continuous with the pericardial sac A prolongation of the pericardial sac extended beneath the xiphoid into the subcutaneous tissues of the anterior abdominal wall The anterior portion of the diaphragm was not attached to the xiphoid or to the costal cartilages at the mid line The musculature of the upper anterior abdominal wall was missing and the finger like projection of the diverticulum extended from the left ventricle through the defect In addition there was a small nodule of myocardial muscle found free in the subcutaneous fat of the upper abdominal wall

The heart was enormously enlarged the enlargement was due solely to the greatly dilated right auricle and the greatly hypertrophied right ventricle The superior and inferior venae cavae entered normally into the right auricle The foramen ovale was anatomically patent The tricuspid valve was normal and opened into a greatly hypertrophied right ventricle The wall of the right ventricle was 1 cm in thickness in many places There was a large defect in the ventricular septum The pulmonary valve and pulmonary ring were normal The pulmonary artery was greatly dilated The pulmonary veins entered a small left auricle, the mitral valve and the left ventricle were normal in size Extending downward from the apex of the left ventricle there was a diverticulum consisting of a long, narrow chamber The orifice of the chamber was only 3 mm in diameter The chamber measured 6 5 cm in length and varied in width as shown in Figure VIII-8 The myocardium of the chamber was thin, it measured approximately 1 to 2 mm in thickness except at the tip There was a high ventricular septal defect The aorta, however, arose normally from the left ventricle and did not over ride the defect The ascending aorta measured 1.2 cm in diameter The left common carotid artery arose anomalously from the innominate artery The aorta between the left subclavian artery and the point of entrance of the ductus arteriosus was only 7 mm in diameter Beyond the ductus arteriosus the aorta again increased to 9 mm in diameter

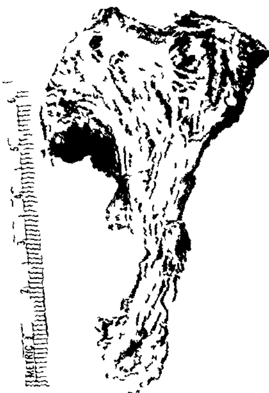


FIGURE XIII-8 Diverticulum of the left ventricle
Case XIII-1

The lungs were grossly normal Microscopic sections of the lungs showed extreme muscular hypertrophy of the smallest pulmonary arterioles with tiny lumina No intimal lesions were seen Thus the small pulmonary arterioles appeared similar to systemic arterioles The diverticulum showed normal endocardium and myocardium The liver was normal

Final anatomical diagnosis Congenital malformation of the heart ventricular septal defect, patent foramen ovale, patent ductus arteriosus and diverticulum of the left ventricle coarctation of the aorta proximal to the ductus arteriosus origin of the left common carotid artery from the innominate artery pulmonary arteriosclerosis right ventricular hypertrophy congenital defect in the pericardium congenital mal formation of the diaphragm diastasis recti abdominis with ventral hernia containing cardiac diverticulum myocardial fibers in abdominal subcutaneous tissue ventral surgical incision with attempted removal of cardiac diverticulum

Comment The diagnosis of diverticulum of the left ventricle was correct The severity of the pulmonary hypertension undoubtedly contributed to the cardiac difficulty and increased the risk of operation, though it was not the apparent cause of death

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CHAPTER XIV

TRUNCUS ARTERIOSUS AND HEMI-TRUNCUS ARTERIOSUS

A TRUNCUS arteriosus means that there is a single great vessel which directs the blood to the systemic circulation and to the lungs "Hemi truncus arteriosus" is the name given to the condition in which one pulmonary artery arises directly from the aorta, as it does in a "true" truncus arteriosus, and the other pulmonary artery arises normally from the right ventricle The former condition is discussed in Section A, the latter in Section B

A Truncus Arteriosus

A truncus arteriosus is a single great vessel of abnormally large caliber which combines the features of both great vessels This vessel receives all the blood from both ventricles and directs the blood to the systemic circulation and to the lungs The circulation to the lungs is by way of the pulmonary artery which arises directly from the base of the single great vessel, or there are no pulmonary arteries and the circulation to the lungs is established through the bronchial arteries or some anomalous vessels of the collateral circulation which may be given off from almost any portion of the aorta The coronary arteries arise from the base of the common trunk, which continues as the aorta and supplies the systemic circulation in the normal manner

Formerly, this malformation was classified as a "true truncus arteriosus" only provided that the orifice was guarded by a valve with four semilunar cusps and pulmonary arteries were given directly off the single great vessel The condition was termed a "pseudo truncus arteriosus" or a "truncus aorticus" if a vestigial pulmonary artery was found, or if the valve guarding its orifice had less than four cusps, even though there was but a single great vessel and the circulation to the lungs was by way of the bronchial arteries The occurrence of four semilunar cusps has been generally discarded as an essential criterion Humphreys,[1] in a careful analysis of an extensive series of cases expressed the opinion that a pseudo truncus arteriosus represents an earlier developmental arrest in the formation of the great vessels than does a "true" truncus arteriosus In a "true" truncus arteriosus the pulmonary artery has developed but has failed to become separated from the aorta in a pseudo truncus arteriosus the pulmonary artery has not yet reached the stage at which it joins the truncus

A review of the development of the great vessels from the primitive aortic arches is necessary to understand the relation of the two conditions and the fundamental nature of the resulting malformation

EMBRYOLOGY

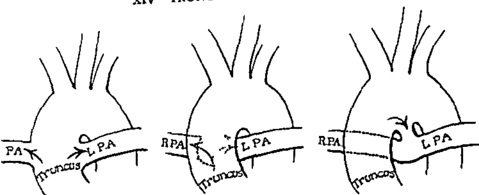
The aorta is derived from the fourth branchial arch and the pulmonary artery from the sixth Normally the sixth left branchial arch distal to the pulmonary artery persists as the ductus arteriosus

Both the fourth branchial arch, which forms the aorta, and the sixth arch, which forms the pulmonary artery, originate from the aortic sac, whence the aortic trunk leads to the bulbus cordis

A so-called "true" truncus arteriosus occurs if the aortic septum fails to develop within the primitive aortic sac, under such conditions the pulmonary artery arises directly from the common trunk Often the only remnant of the aortic septum present is a fold in the wall of the aorta just above the point where the main pulmonary artery branches off from the common trunk In other cases the formation of the aortic septum has progressed to the stage at which the left and right pulmonary arteries arise from the single great vessel through a common orifice This common orifice or common vessel may be given off from the posterior wall of the ascending aorta or higher up on the arch of the aorta (see the diagram of the truncus arteriosus in Figure XIV–1) In the rare instances in which the pulmonary arteries arise directly from the truncus arteriosus, the ductus arteriosus is normally formed, in most instances, however, it is absent Inasmuch as the function of the ductus arteriosus is to direct the blood from the pulmonary artery to the aorta, it serves no purpose when the pulmonary artery and the aorta arise from a common trunk Therefore, since the ductus arteriosus is of no functional importance, it is not surprising to find that it may atrophy and disappear quite as readily as does the distal end of the sixth right branchial arch

The condition is also considered a "true" truncus arteriosus when there is a single large truncus guarded by a valve with four semilunar cusps and no vestige of a pulmonary artery is found Under such circumstances the circulation to the lungs is through the bronchial arteries or other pathways of the collateral circulation Such vessels are usually minute and the pulmonary blood flow is meager

A pseudo truncus arteriosus represents an arrest in the formation of the sixth branchial arch which is of such a nature that it fails to meet the aortic sac Usually, when the proximal end of the sixth branchial arch fails to meet the aortic



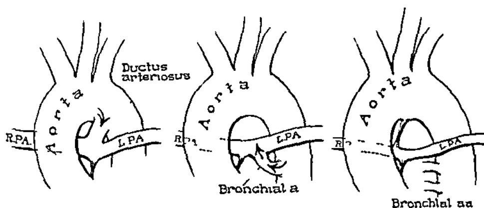


FIGURE XIV–1 Truncus arteriosus with pulmonary vessels arising from the truncus and pseudo truncus arteriosus

sac, the distal end fails to meet the dorsal aorta Consequently, not only does the pulmonary artery fail to meet the aorta in the normal fashion, but the ductus arteriosus is absent Under such circumstances a vestigial pulmonary artery exists, it leads to the lungs but does not connect with the heart or the aorta and consequently it carries no blood Under such circumstances, as in a true truncus arteriosus in which the pulmonary artery is absent, the only way for the blood to reach the lungs is by way of the collateral circulation, which is usually established through the bronchial arteries Thus the circulation to the lungs may be identical in the two conditions

Humphreys showed that in a very real sense a true truncus arteriosus and a pseudo truncus arteriosus could exist together She cited a case in which one pulmonary artery arose directly from the aorta and the other pulmonary artery

the coronary arteries has not been seen by *the author*, either at operation or at autopsy, or at least it has *not* been recognized as such

Although in most instances, it is relatively easy *to* locate the anomalous vessels as they arise from the aorta *or* its branches, it is extremely *difficult to trace* their entrance into the lungs Occasionally, as *mentioned above*, a large vessel can be seen to enter the pulmonary artery In most instances, *however*, it is *well* nigh impossible to demonstrate the exact manner *in which the* vessels of collateral circulation *anastomose* with the pulmonary vascular bed

Collister et al [4] have made careful studies of the development of the pulmonary vascular bed They believe that when there is pulmonary atresia the pathways of the collateral circulation are laid down during intra uterine life *This* must certainly be true in the case of a truncus arteriosus in which the entire circulation to the lungs is by way of anomalous vessels If it is true in one instance, it is certainly reasonable to believe that it is true in another The concept implies that the ultimate effectiveness of the collateral circulation is dependent upon the vascular channels laid down during intra uterine life

It has been the author s experience that some patients with a pseudo truncus arteriosus have been greatly helped by a systemic pulmonary anastomosis and that other patients have developed thromboses far out in the lungs distal to the anastomosis Thus it appears that in some instances the pulmonary arteries are able to direct blood to the lungs and that in other instances the vestigial pulmonary artery does not open normally into the pulmonary vascular bed It is the author s impression that if the vessels of the collateral circulation open into the pulmonary arteries, then the distal pulmonary vascular bed is normal and can direct the blood to the lungs, if, however, the vessels of the collateral circulation enter the lungs far out in the capillary bed, *or* connect with the bronchial arterial system then the vestigial pulmonary artery lacks its normal connection with the pulmonary capillary bed and at best it can carry only a small volume of *blood to the lungs*

Furthermore even upon direct observation at operation, it may be impossible to determine whether or not there is a vestigial pulmonary artery Consequently the differentiation of a true truncus arteriosus from a pseudo truncus arteriosus, on the basis of the existence of a *rudimentary* pulmonary artery, can be established only at autopsy Because of this and because there are all gradations of both types of truncus arteriosus, these two conditions are *considered as* variants of the same malformation namely a truncus arteriosus

Additional evidence that these two conditions represent closely *related* de

was a blind tube, that is, it failed to connect with the heart or the aorta, the circulation to the lung with the blind pulmonary artery was established by way of the bronchial arteries The author has seen two cases in which one pulmonary artery arose from the aorta and the other arose in the normal fashion from the right ventricle (see Section B)

Christeller,[2] in his detailed study of a series of cases of pulmonary atresia, found that when the ductus arteriosus did not develop or did not remain patent there were eight other possible pathways by which the collateral circulation might be established

1 The anterior bronchial arteries, that is, the superior bronchial arteries which arise from the arch of the aorta
2 The posterior bronchial arteries, which arise from the posterior wall of the descending aorta or from the intercostal arteries
3 The anterior mediastinal arteries, which arise from the internal mammary arteries
4 The posterior mediastinal arteries which arise from the posterior wall of the aorta
5 Branches from the esophageal arteries, which arise from the posterior mediastinal arteries
6 Branches from the pericardial arteries which arise either from the esophageal arteries or from the posterior mediastinal arteries
7 Anomalous branches which may arise from the coronary arteries of the heart
8 Anomalous arteries which may arise from the aorta or occasionally from the subclavian arteries, from the diaphragmatic arteries, or from the pericardial arteries

Enlargement of the anterior and posterior mediastinal vessels has been commonly observed at operation In almost every instance the collateral circulation over the anterior chest wall is evident and the mediastinal vessels are large and numerous Furthermore, the posterior mediastinal vessels may be so numerous as to cause increased hilar shadows In many patients large anomalous vessels coursing across the lungs have been seen at operation In one instance a large vessel which apparently arose from the descending aorta opened directly into the inferior wall of the pulmonary artery, as shown in the middle diagram of pseudo truncus arteriosus in Figure XIV–1 Dr Paul White[3] has seen one patient in whom at operation the circulation to the left lung apparently came from a large vessel which arose from the descending aorta The author has seen one similar instance in which at autopsy the main vessel of collateral circulation was found to arise from the aorta below the level of the diaphragm In addition, she has also seen one child who suffered from hematemesis which, in all probability, was due to hemorrhage from esophageal varices secondary to the enlargement of the esophageal arteries Collateral circulation from the pericardial arteries or

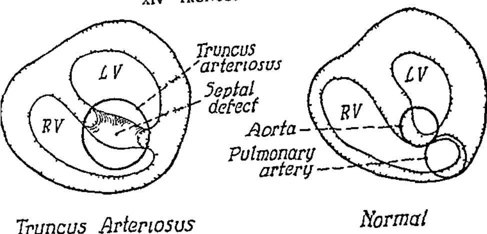


FIGURE XIV-2 Truncus arteriosus and normal heart

ductus arteriosus is usually absent when present, it is seldom of functional importance

A truncus arteriosus is often associated with an arrest in the development of the heart itself Defects in the auricular septum are common, occasionally the ventricular septum fails to develop and there is but a single ventricle Such conditions, however, represent additional anomalies, they are not part and parcel of the truncus arteriosus A single auricle or even a single ventricle does not greatly alter the circulation, because all the blood is pumped out into the common vessel Therefore, regardless of whether or not venous and arterial blood are mixed in the auricle or in the ventricle, complete admixture occurs in the truncus arteriosus Furthermore, there is no separation of the work of the two sides of the heart Either a common ventricle or two ventricles pump the blood through a single great vessel to both the systemic and the pulmonary circulation

COURSE OF THE CIRCULATION

During fetal life the circulation to the lungs is minimal, the main flow of blood is by way of the single vessel to the body of the fetus The blood is returned in the normal manner by the superior vena cava and the inferior vena cava to the right auricle The blood which goes to the lungs is returned in the normal fashion to the left auricle Nevertheless, inasmuch as the truncus arteriosus arises more from the right ventricle than from the left, the work of the right ventricle is increased (see Figures XIV-7 and 8)

After birth the establishment of respiration opens the pulmonary vascular bed and thereby some blood from the truncus arteriosus is directed to the lungs

velopmental arrests is that they cause virtually the same alteration in the size and the shape of the heart The fundamental contour of the heart is determined during intra uterine life During fetal life the circulation to the lungs is relatively unimportant, consequently the work of the heart is virtually the same whether the circulation to the lungs is from the pulmonary artery or from the bronchial arteries Moreover, when the circulation to the lungs is by way of the bronchial arteries, existence of a rudimentary pulmonary artery is of no functional importance In both cases, the two ventricles pump the blood through the single great vessel to the body of the fetus

Moreover, in either a pseudo truncus arteriosus or a "true" truncus arteriosus, the pulmonary blood flow may be excessive or it may be markedly reduced Therefore, instead of an anatomical separation of the two conditions, the author prefers a physiological differentiation on the basis of the volume of the pulmonary blood flow, which may be increased, normal, or decreased

At the stage in the development of the heart when there is but a single great vessel, there are but a single auricle and a single ventricle, the single great vessel arises from the primitive bulbus cordis As the aortic septum develops, the common trunk shifts from its position relative to the bulbus cordis to assume a more posterior position above the left ventricle When the development of the great vessel is arrested, the truncus arteriosus arises from both ventricles, but usually more from the right than from the left

NATURE OF THE MALFORMATION

A truncus arteriosus is a single great vessel of unusually large caliber which combines the features of both great vessels The common truncus over rides the ventricular septum and receives the blood from both ventricles, hence a high ventricular septal defect is an integral part of the malformation (see Figure XIV–2) The orifice of the truncus is guarded by a valve with two, three, or four semilunar cusps The coronary arteries arise from the base of the great vessel The pulmonary artery arises either from the posterior wall of the truncus (see Figure XIV–3) or from its base as in the case reported by MacGilpin,[6] which is shown in Figures XIV–4 and 5, or else the pulmonary artery is absent and the circulation to the lungs is established by way of the bronchial vessels or by some anomalous vessels as illustrated in Figure XIV–6 In all instances the blood from both ventricles is pumped out into the common arterial trunk and thence it flows through the arterial pathways to the systemic circulation and to the lungs The heart is slightly to moderately enlarged Both ventricles are thick walled The



FIGURE XIV 4 Truncus arteriosus with a single pulmonary artery arising from the base of the truncus (same patient as in Figures XIV–3 18)

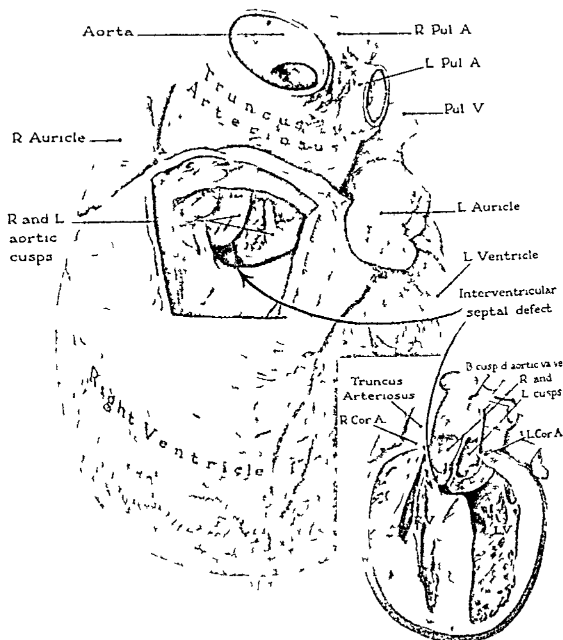


FIGURE XIV-3 Truncus arteriosus with the pulmonary arteries arising from the main truncus and a high ventricular septal defect

The single vessel over rides the septum and receives blood from both ventricles The left and the right coronary arteries arise from the base of the aorta behind the semilunar cusps



FIGURE XIV 4 Truncus arteriosus with a single pulmonary artery arising from the base of the truncus (same patient as in Figures XIV–5 18)

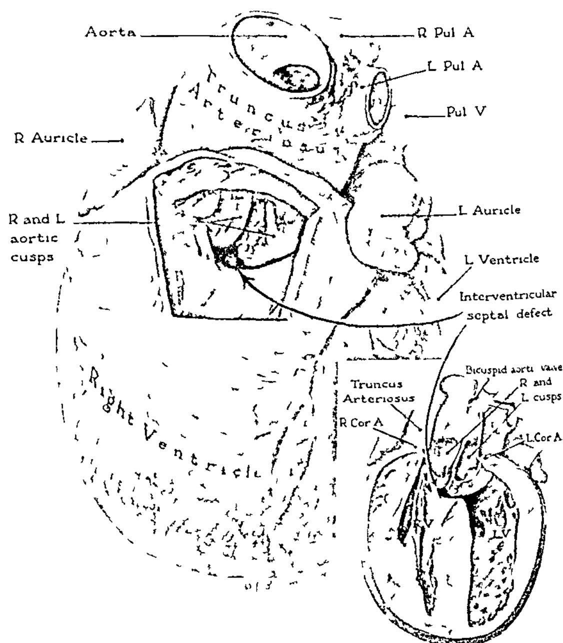


FIGURE XIV-3 Truncus arteriosus with the pulmonary arteries arising from the main truncus and a high ventricular septal defect

The single vessel over rides the septum and receives blood from both ventricles The left and the right coronary arteries arise from the base of the aorta behind the semilunar cusps

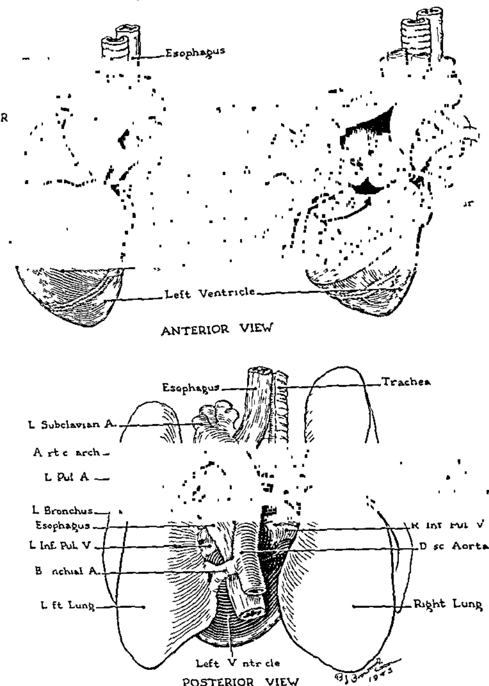


FIGURE XIV–6 Pseudo truncus arteriosus (same patient as in Figure XIV–20)

The anterior view shows the truncus arteriosus over riding the ventricular septum The posterior view shows the enlarged bronchial arteries and the esophagus caught between the bronchial arteries and displaced to the left

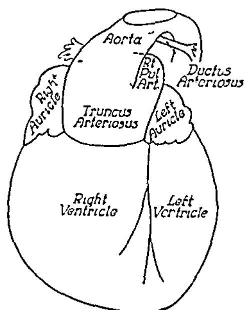


FIGURE XIV-5 Truncus arteriosus with a single pulmonary artery arising from the base of the truncus (same patient as in Figure XIV-4)

If the pulmonary arteries arise from the truncus, a large volume of blood is directed to the lungs, where it is oxygenated, and returned in the normal manner by the pulmonary veins to the left auricle, thence it flows to the left ventricle The remainder of the blood in the truncus arteriosus is directed to the body and is returned by the superior and inferior venae cavae to the right auricle, thence it flows to the right ventricle The blood from both ventricles is pumped out into the common trunk There the cycle starts again Under such circumstances a large volume of oxygenated blood is mixed with the venous blood which is returned from the body to the right side of the heart, therefore cyanosis is minimal or absent (see Diagram XIV-1)

When the pulmonary artery fails to meet the aorta and the ductus arteriosus is absent, the only possible way for blood to reach the lungs is through the bronchial arteries or some anomalous vessels Although these pathways of collateral circulation enlarge, they are seldom sufficient to permit adequate circulation to the lungs The volume of blood which flows to the lungs varies with the size and number of these pathways When these are few or of very small caliber, the pulmonary blood flow is extremely meager Only a small volume of oxygenated blood is returned to the left auricle and the left ventricle Hence only a small volume of oxygenated blood is mixed with a large volume of venous blood, consequently cyanosis is intense (see Diagram XIV-2)

Thus a truncus arteriosus may have excessive, normal, or reduced pulmonary blood flow Nevertheless, the course of the circulation in all instances is basically the same

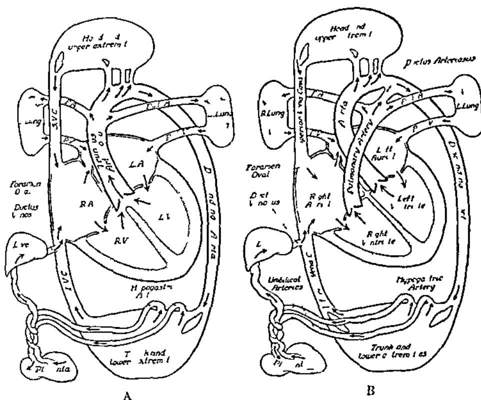


FIGURE XIV-8 Fetal circulation (A) Truncus arteriosus with the pulmonary arteries arising from the main truncus and (B) normal heart

nary artery is related to volume of the pulmonary blood flow When relatively small vessels are given off the descending aorta, the pulmonary pressure is usually abnormally low

CLINICAL FINDINGS

The presence or absence of cyanosis varies with the adequacy of the pulmonary blood flow Cyanosis is minimal or absent in infancy and childhood when the pulmonary arteries arise directly from the common trunk Under such circumstances a large volume of blood reaches the lungs for oxygenation and a large volume of oxygenated blood is returned to the left side of the heart Furthermore, the blood is directed into the pulmonary arteries under systemic pressure Consequently, if the pulmonary vascular bed opens up in the normal fashion, there is real danger of excessive flow to the lungs and the depletion of the systemic circulation This phenomenon is the probable explanation of the high mortality rate in the neonatal period when the pulmonary arteries arise

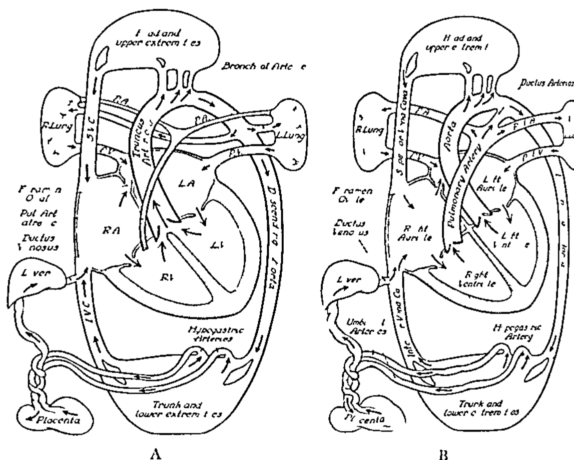


FIGURE XII–7 Fetal circulation (A) Truncus arteriosus with circulation to the lungs by way of the bronchial arteries and (B) normal heart

PHYSIOLOGY OF THE MALFORMATION

In this malformation all the blood to the body and to the lungs is pumped out through the common truncus Consequently the same admixture of venous and arterial blood is directed to the two circulations The height of the pressure in the lesser circulation depends upon the size and the position of the vessels which direct the blood to the lungs The pressure in the ascending aorta is a combination of the force of the actual expansion of the aorta and that of the for ward propulsion of the blood, whereas in the descending aorta the expansile force is no longer present and the pressure is caused solely by the force of the forward propulsion of the blood [7, 8] Consequently, if large pulmonary arteries are given off the ascending portion of the truncus arteriosus, the pressure in the pulmonary artery is the same as that in the systemic circulation, under such cir cumstances pulmonary hypertension is severe If, however, large vessels are given off the descending aorta, the hemodynamics are similar to those which occur with a huge ductus arteriosus, namely, the height of the pressure in the pulmo-

Diagram XIV–1

Truncus arteriosus with the pulmonary arteries arising from the main truncus

The essential feature of this malformation is that there is but a single great vessel which combines the features of both great vessels, it receives the blood from both ventricles and directs blood to the systemic circulation and to the lungs The coronary arteries arise from the base of this single vessel and the pulmonary arteries originate as branches of the main trunk Both auricles are normally formed and both ventricles are normally formed except for a high ventricular septal defect This is inevitable as the truncus arteriosus arises from both ventricles, hence it over rides a high ventricular septal defect Both ventricles are abnormally thick

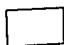
The blood from the right auricle flows into the right ventricle and that from the left auricle flows into the left ventricle. From both ventricles the blood is pumped out into the common arterial trunk and flows both to the body and through the pulmonary arteries to the lungs The blood from the head and the upper extremities is returned by the superior vena cava to the right auricle and that from the trunk and the lower extremities is returned by the inferior vena cava to the right auricle The blood which is pumped into the pulmonary arteries flows in the normal manner to the lungs where it is oxygenated and the oxygenated blood is returned by the pulmonary veins to the left auricle There the cycle starts again Inasmuch as the pulmonary arteries are of normal size a large volume of oxygenated blood is returned to the left auricle and to the left ventricle This blood is pumped out into the common arterial trunk where it mixes with venous blood from the right side of the heart Inasmuch as a large volume of oxygenated blood is mixed with the normal venous blood, cyanosis is minimal or absent.

Clinical diagnosis If the pulmonary arteries are large and are given off at the base of the truncus the pressure in the pulmonary arteries is the same as that in the aorta there is marked pulmonary hypertension and consequently there is no continuous murmur

Frequently both pulmonary arteries are given off from a single orifice, which creates a mild functional pulmonary stenosis under such circumstances either an early diastolic murmur is audible at the base of the heart or a continuous murmur is audible over the lungs The electrocardiogram usually shows a right axis deviation and evidence of either right ventricular hypertrophy or combined ventricular hypertrophy

DIAGRAM XIV–1

Head and upper extremities
R Lung
L Lung
S.V.C.
P.A.
P.V.
Pul A.
Pul V
Truncus Arteriosus
L.A
R.A.
L.V
R.V.
Descending Aorta
Coronary-Artery
I.V.C.
Trunk and lower extremities


Arterial blood (fully saturated)


Small admixture of venous blood
No visible cyanosis


Venous and arterial blood
Cyanosis visible


Venous blood

Diagram XIV–1

Truncus arteriosus with the pulmonary arteries arising from the main truncus

The essential feature of this malformation is that there is but a single great vessel which combines the features of both great vessels, it receives the blood from both ventricles and directs blood to the systemic circulation and to the lungs The coronary arteries arise from the base of this single vessel and the pulmonary arteries originate as branches of the main trunk Both auricles are normally formed and both ventricles are normally formed except for a high ventricular septal defect This is inevitable as the truncus arteriosus arises from both ventricles, hence it over rides a high ventricular septal defect Both ventricles are abnormally thick

The blood from the right auricle flows into the right ventricle and that from the left auricle flows into the left ventricle. From both ventricles the blood is pumped out into the common arterial trunk and flows both to the body and through the pulmonary arteries to the lungs The blood from the head and the upper extremities is returned by the superior vena cava to the right auricle and that from the trunk and the lower extremities is returned by the inferior vena cava to the right auricle The blood which is pumped into the pulmonary arteries flows in the normal manner to the lungs where it is oxygenated and the oxygenated blood is returned by the pulmonary veins to the left auricle There the cycle starts again Inasmuch as the pulmonary arteries are of normal size a large volume of oxygenated blood is returned to the left auricle and to the left ventricle This blood is pumped out into the common arterial runk where it mixes with venous blood from the right side of the heart Inasmuch as a large volume of oxygenated blood is mixed with the normal venous blood, cyanosis is minimal or absent.

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Diagram XIV-2

Head and upper extremities

Pulmonary Arteries

Bronchial Arteries

R Lung

S.V.C.

PV

Pseudo-Truncus Arteriosus

L Lung

Pul V

L.A

Descending Aorta

R.A

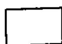
Remnant of Pul Art

L.V

R.V

I.V.C.

Trunk and lower extremities

 Arterial blood (fully saturated)

 Venous and arterial blood
Cyanosis visible

 Small admixture of venous blood
No visible cyanosis

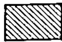
 Venous blood

DIAGRAM XIV-2

Pseudo truncus arteriosus

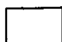
In this malformation there is but a single great vessel which receives the blood from both ventricles and directs it to the systemic circulation and to the lungs The pulmonary arteries, however do not arise from this *great vessel* The circulation to the lungs is by way of the bronchial arteries or anomalous vessels of the collateral circulation Both auricles and both ventricles are normally formed, inasmuch as the common arterial trunk arises from both ventricles a high ventricular septal defect is inevitable The coronary arteries arise at the base of the truncus arteriosus behind the semilunar cusps Inasmuch as the pulmonary artery *does not connect* with the truncus arteriosus or with the heart the only pathway by which the blood can reach the lungs is through the bronchial arteries There may be one or two abnormal superior bronchial arteries which arise from the arch of the aorta or some of the posterior bronchial arteries may be abnormally large

The blood from the right auricle flows into the right ventricle and is pumped out by way of the common arterial trunk to the body and through the bronchial arteries to the lungs The blood from the head and the upper extremities is returned by the superior vena cava to the right auricle and that from the trunk and the lower extremities is returned by the inferior vena cava to the same auricle The blood in the bronchial arteries flows to the lungs and the oxygenated blood is returned in the normal manner by the pulmonary veins to the left auricle Thence the blood flows to the left ventricle and is pumped out into the truncus arteriosus There the cycle starts again

Clinical diagnosis The patient usually shows persistent cyanosis but is not severely limited The heart is enlarged there is a concave curve at the base of the heart to the left of the sternum a prominent high aortic knob and decreased hilar markings, and an absence of the hilar comma A continuous murmur is audible over one or both lungs the intensity of the murmur is inversely proportional to the intensity of the cyanosis The electrocardiogram usually shows a right axis deviation and commonly shows evidence of combined hypertrophy

DIAGRAM XIV–2

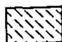
Head and upper extremities
Pulmonary Arteries
Bronchial Arteries
R Lung
S.V.C.
PV
Truncus Arteriosus
Pul V
L Lung
LA
Descending Aorta
R.A.
Remnant of Pul Art
LV
R.V.
I.V.C.
Trunk and lower extremities



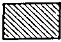
Arterial blood (fully saturated)



Venous and arterial blood
Cyanosis visible



Small admixture of venous blood
No visible cyanosis



Venous blood

become obliterated; hence most infants with a truncus arteriosus do not suffer from attacks of paroxysmal dyspnea Moreover, the vessels of collateral circulation tend to increase in size as the patient grows and consequently the circulation to the lungs may gradually show slight improvement

Exercise tolerance of the individual varies directly with the volume of the pulmonary blood flow When the pulmonary arteries arise *directly from* the aorta, there is adequate or excessive pulmonary blood flow and *the individual* can do virtually as much as any normal person In contrast to this, when the circulation to the lungs is by way of the minute vessels of collateral circulation, *the* patient s activity is extremely limited An intermediate stage between these two extremes is the most common of all Namely, the collateral circulation is moderately well developed Under such circumstances, although the pulmonary *blood flow is reduced, it is constant* Furthermore, since the arteries which carry *the blood to the lungs arise directly from the aorta*, any increase in the systemic pressure increases the circulation to the lungs The result is that most patients can walk considerable distances and do not squat when tired In this malformation the systemic pressure regulates the pressure under which the blood flows to the lungs Indeed, the pulmonary blood flow remains relatively constant unless there is a collapse of the systemic circulation It is important to remember that in a patient whose pulmonary blood flow is extremely meager a sudden drop in the systemic pressure may be fatal

The pulse is forceful and of equal strength in the arm and the leg Arterial pulsations are frequently conspicuous in the vessels *of the neck*

The blood pressure is, however, usually normal Indeed, the diastolic pressure *is usually higher* than might be expected from the quality of the pulse

CARDIAC FINDINGS

The heart is slightly to moderately enlarged The enlargement mainly involves the right ventricle which may be so great as to cause left sided chest deformity

The second sound at the base is loud and pure It may be better heard to the left of the sternum than to the right it is however never reduplicated

A harsh systolic murmur is frequently audible over the *precordium and* may be widely transmitted throughout the *chest When the murmur* is intense, a *thrill* develops at an early age the thrill is of maximal intensity over the sternum at the base of the heart

A continuous murmur which is best heard over the lungs is the most charac

directly from the base of the truncus Once a balance is established, the patient usually does well for a number of years Unfortunately when the pulmonary arteries arise from the base of the truncus, these vessels receive blood under systemic pressure As in all such instances, initially the pulmonary bed opens up slowly and the hypertension is compensatory Over the years, however, intimal changes occur and become progressively more severe Eventually the vascular changes may be so great as to reduce the volume of blood which reaches the lungs for oxygenation and thus they increase the volume of venous blood directed to the systemic circulation to such an extent that the patient develops cyanosis and polycythemia Therefore, terminally, most adults with this malformation show varying degrees of cyanosis, clubbing, and polycythemia

Cyanosis almost always dates from birth when the pulmonary artery is absent and the circulation to the lungs is through the bronchial arteries or anomalous vessels of the collateral circulation Only rarely are these vessels sufficiently large so that there is adequate circulation to the lungs Usually these pathways are extremely small in comparison with the size of the normal pulmonary artery and the pulmonary blood flow is proportionately reduced Under such circumstances, only a small volume of blood goes to the lungs and a large volume of blood goes to the body, hence only a small volume of oxygenated blood is returned to the left auricle and a large amount of venous blood is returned to the right auricle. Regardless of whether or not there is a gross defect in the auricular septum, all the blood returned to the auricles is pumped out into the common great vessel, hence the greater the reduction of the pulmonary blood flow, the smaller is the volume of oxygenated blood directed to the systemic circulation and the lower is the oxygen saturation of the arterial blood Under such circumstances cyanosis is readily apparent at birth and polycythemia develops in early infancy

Dyspnea varies with the adequacy of the pulmonary blood flow Nevertheless, the volume of the pulmonary blood flow, regardless of whether it is great or small, is related to the systemic pressure, therefore it is relatively constant For this reason, although polypnea may be marked, respirations are not labored and attacks of paroxysmal dyspnea are rare Indeed, paroxysmal dyspnea occurs only in those rare instances in which the ductus arteriosus constitutes the principal pathway by which the blood can reach the lungs As the ductus periodically constricts during the process of closure, the circulation to the lungs is abruptly reduced, under such circumstances, an infant may suffer from attacks of paroxysmal dyspnea Usually, however, the circulation to the lungs is maintained by small arteries which arise directly from the aorta These vessels do not tend to

ter heard posteriorly than anteriorly The murmur may be maximal over the base of the heart to the left of the sternum, under such circumstances it requires differentiation from the murmur of persistent patency of the ductus arteriosus The murmur produced by a truncus arteriosus differs from that associated with patency of the ductus arteriosus in that it has a humming quality and varies in intensity with respiration, it is usually better heard with the breath held in full expiration Ordinarily the murmur is not very loud and is limited to one side of the chest It may be localized in almost any place, depending on the location of the anomalous vessels to the lungs Thus the murmur may be audible over a limited area, front, back, or in the axilla Such a murmur frequently escapes detection unless carefully sought for when listening over the lungs

A thrill is palpable only when the murmur is excessively loud When a thrill does occur it is not localized over the pulmonary area but is widely transmitted throughout the lungs and may be readily palpable in both axillae A continuous thrill, however is the exception not the rule

X RAY AND FLUOROSCOPIC FINDINGS

In infancy the contour of the heart is distinctive, especially when the pulmonary arteries are absent In the anterior posterior position the heart is seen to be greatly enlarged Owing to the absence of the normal pulmonary artery, the shadow cast by the pulmonary conus is absent the upper margin of the cardiac shadow to the left of the sternum is concave The ventricular shadow extends nearly to the axilla and the apex of the heart is upturned owing to the enlargement of the right ventricle (see Figures xiv–9 and 10)

In the right anterior-oblique position the upper margin of the shadow cast by the right ventricle makes an abrupt angle with the aorta and extends horizontally outward to the anterior chest wall It is so straight that it has the appearance of a shelf or wall (see Figures xiv–11 and 12) If Puck could sit on the edge of a non functioning right ventricle, Humpty Dumpty could balance on this wall

In the left anterior-oblique position the shelf may or may not be apparent, depending on the thickness of the right ventricle Inasmuch as the truncus arises more from the right ventricle than from the outflow tract, there is usually an angulation between the truncus arteriosus and the right ventricle In Figure xiv–11 the patient is not rotated sufficiently far to show the enlargement of the right ventricle but the anterior displacement of the esophagus by the retro-esophageal vessels is clearly visible (see below)

teristic of all findings in a truncus arteriosus The murmur is seldom present at birth but frequently develops during the early months of life It is dependent on the continuous flow of blood from the aorta through the vessels of the collateral circulation to the lungs Thus it depends on the height of the systemic pressure, on the size of the vessel leading to the lungs, and on the pulmonary resistance

When the pulmonary arteries arise directly from the base of the heart, the pressure in these vessels is the same as that in the systemic circulation If the lungs expand rapidly, the pulmonary blood flow becomes excessive, when this occurs no continuous murmur is heard and the infant dies at an early age Often the orifice of the pulmonary artery is sufficiently small to break the high pressure in the truncus Under such circumstances, as the pulmonary vascular bed gradually opens, the difference in pressure between the systemic and the pulmonary circulation becomes sufficiently great to cause a continuous murmur

If the vessels of the collateral circulation are minute, so little blood flows through these vessels that there will be no continuous murmur Fortunately, the vessels of the collateral circulation tend to increase in size with the growth of the individual Therefore it is not uncommon for a continuous murmur to appear during infancy As previously mentioned, in rare instances when the main pathway of the collateral circulation is by way of the ductus arteriosus, as the ductus arteriosus undergoes obliteration the infant may suffer from attacks of paroxysmal dyspnea The author has had the opportunity to examine two such patients during episodes of paroxysmal dyspnea In both instances the continuous murmur disappeared during the attack of paroxysmal dyspnea and upon the return of the murmur the infant s color immediately improved and dyspnea subsided

Usually, however, the vessels are of moderate size—small enough to break the force of the systemic pressure and yet sufficiently large to permit blood to flow continuously to the lungs The intensity of the continuous murmur is directly proportional to the volume of blood which reaches the lungs Consequently, if the patient shows minimal or no cyanosis and correspondingly little incapacity, there is a loud continuous murmur If there is moderate cyanosis and some limitation of activity, the murmur may be localized over one lung or over one portion of one lung, whereas if there is intense cyanosis and marked incapacity, there may be no murmur

The murmur may be of maximal intensity in almost any location If the murmur is loud, it is usually well heard over one or both lungs It is frequently bet

ter heard posteriorly than anteriorly The murmur may be maximal over the base of the heart to the left of the sternum, under such circumstances it requires differentiation from the murmur of persistent patency of the ductus arteriosus *The murmur* produced by a truncus arteriosus differs from that associated with patency of the ductus arteriosus in that it has a humming quality and varies in intensity with respiration, *it is* usually better heard with the breath held in full expiration Ordinarily the murmur is not very loud and is limited to one side of the chest It may be localized in almost any place, depending on the location of the anomalous vessels to the lungs Thus the murmur may be audible over a limited area front, back, or in the axilla Such a murmur frequently escapes de tection unless carefully sought for when listening over the lungs

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FIGURE XIX-9 Truncus arteriosus (same patient as in Figures XIX-11, 21). Infant.

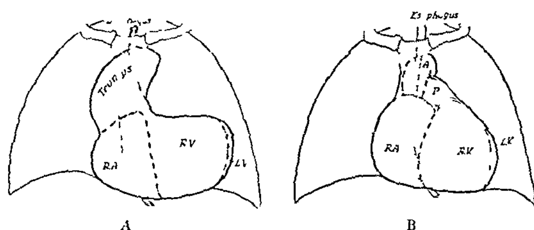


FIGURE XIX-10 (A) Truncus arteriosus and (B) normal heart. Infant.

The truncus arteriosus is of abnormally large caliber and arches at an abnormally high level as shown in Figure XIX-9 and also in Figures XIX-13, 15, 19, and 24. In infancy the abnormally large aorta is not only conspicuous but also usually indents the esophagus. The indentation of the esophagus can generally be demonstrated in the anterior posterior position and in both oblique positions upon its delineation with a radio-opaque mixture. A right aortic arch is quite common with this malformation, as shown in Figures XIX-13 and 14.

As the individual grows, the diaphragm descends and the heart comes to occupy a more vertical position in the chest than it did in infancy. When this



Left anterior-oblique position



Right anterior-oblique position

FIGURE XIV–11 Truncus arteriosus (same patient as in Figure XIV–9) Infant

Note the shelf like projection of the right ventricle

occurs the contour of the heart usually changes The right ventricle no longer projects forward so abruptly, consequently the contour of the heart comes to resemble that of a tetralogy of Fallot with a severe pulmonary stenosis (see Figures XIV–15 and 16) Under such circumstances the high aortic arch, in combination with an absence of the shadow cast by the normal pulmonary artery and its branches, gives the clue to the diagnosis

Absence of the shadow cast by the right and left branches of the main pulmonary artery may be striking Danelius[9] emphasized the absence of a hilar comma in cases of truncus arteriosus In the author s experience, not only is the

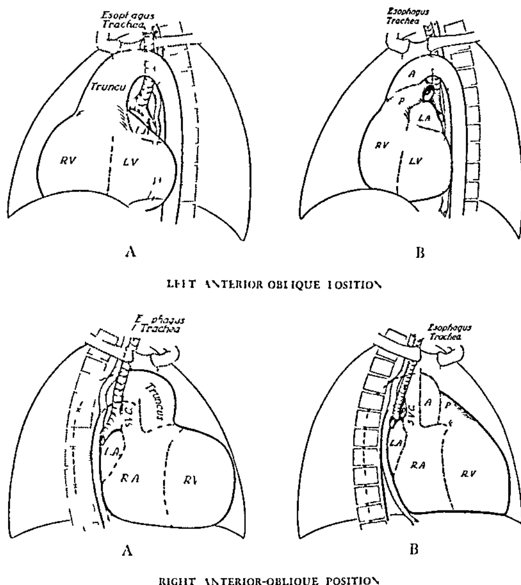


Figure xiv-12 (A) Truncus arteriosus and (B) normal heart Infant

hilar comma absent, but it is replaced by diffuse fine vascular markings which radiate from both hilar regions to the lungs

On the other hand, *if large pulmonary arteries are given directly off from the common trunk,* they receive a large volume of blood under systemic pressure Under such circumstances, although the shadow at the base of the heart has a concave curve, there may be a conspicuous hilar dance (see Figure xiv–17)

Occasionally one or both pulmonary arteries arise from the base of the truncus on the left, close to the origin of the normal pulmonary artery, as shown in Figures xiv–4 and 5 Under such circumstances, in contrast to the usual con

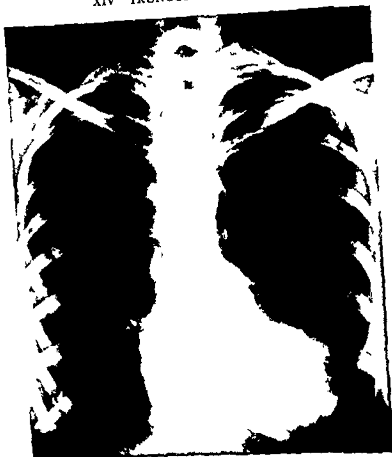


FIGURE XIV-13 Truncus arteriosus with a right aortic arch. Adult

cavity there may be fullness in the region of the pulmonary conus, as shown in Figure XIV-18

The occurrence of anomalous retro-esophageal vessels offers additional evidence that the circulation to the lungs is by way of collateral pathways The esophagus may be caught between anomalous bronchial arteries and displaced anteriorly or laterally as shown in Figures XIV-11 and 12 Indeed, the demonstration of retro-esophageal vessels in the lower thoracic region is of diagnostic significance, as it cannot be due to any abnormality of the vessels which arise from the aortic arch (see Chapter XXVI) In rare instances the esophagus may be caught by one or more of these anomalous vessels and displaced in an extremely bizarre manner as shown in Figure XIV-19

When the main circulation to the lungs is by way of the superior bronchial arteries these vessels may become greatly dilated In rare instances the increased

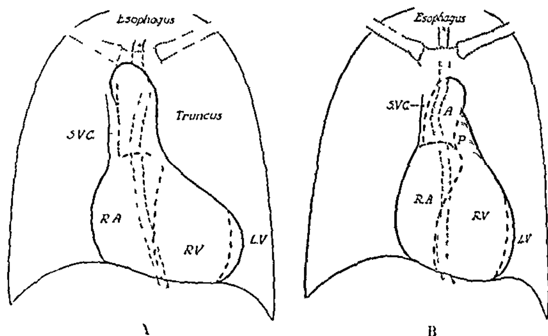


FIGURE XIV–14 (A) Truncus arteriosus with a right aortic arch and (B) normal heart Adult

number of vessels given off the transverse portion of the aorta may cause the aortic arch to be fuzzy and indistinct (see Figure XIV–20)

In summary, there are five findings of diagnostic importance which should be sought for upon radiological examination

1 Moderate cardiac enlargement with a concave curve at the base of the heart to the left of the sternum
2 A prominent aortic knob which lies at an abnormally high level
3 Absence of the main right and left pulmonary arteries
4 Diffuse fine hilar markings
5 Retro esophageal vessels or bizarre displacement of the esophagus

It is, however, important to remember that retro-esophageal vessels are not always present and that the occurrence of a hilar dance does not exclude the possibility of a 'true' truncus arteriosus with the pulmonary artery arising directly from the aorta

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram is subject to considerable variation in most instances the standard leads show a right axis deviation and the precordial leads show evidence of hypertrophy of both ventricles Not infrequently the deflections are of



FIGURE XIV-15 Truncus arteriosus with a left aortic arch Adult

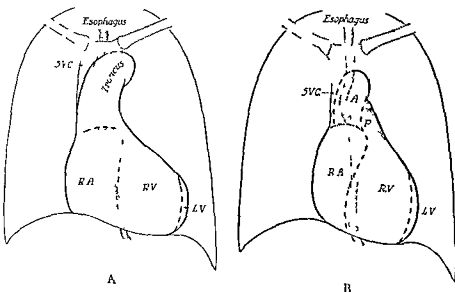


FIGURE XIV-16 (A) Truncus arteriosus with a left aortic arch and (B) normal heart Adult

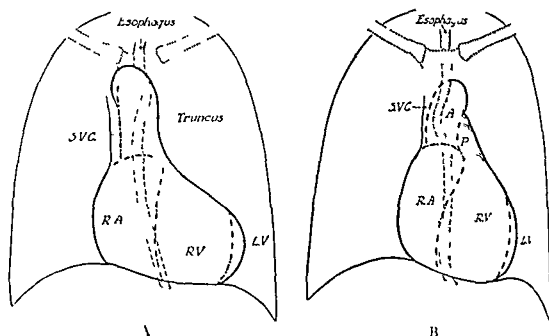


Figure XIV–14 (A) Truncus arteriosus with a right aortic arch and (B) normal heart Adult

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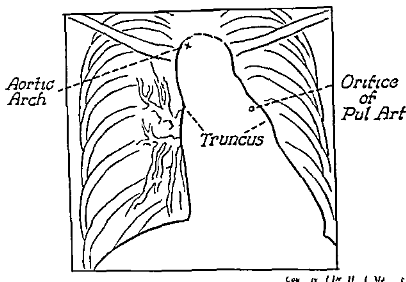


FIGURE XIV 18 Truncus arteriosus with a single pulmonary artery arising from the base of the truncus (tracing of x ray) (same patient as in Figure XIV 4)



FIGURE XIV 19 Truncus arteriosus Child

Note the displacement of the esophagus by retro-esophageal vessels



FIGURE XIV–17 Truncus arteriosus with large pulmonary arteries arising from the truncus Child

abnormally great amplitude (see Figure XIV–21) but this finding is not of diagnostic significance Occasionally V_1 shows evidence of right ventricular hypertrophy (see Figure XIV–22), in rare instances the deflection in V_1 is primarily downward (see Figure XIV–23)

SPECIAL TESTS

The circulation time is abnormally short When the pulmonary arteries arise directly from the aorta, the test material may be so rapidly dissipated into the two circulations that a satisfactory end point is not obtainable

The oxygen saturation of the arterial blood varies inversely with the adequacy of the pulmonary blood flow Even though the pulmonary blood flow is adequate or excessive, since blood from both ventricles is directed into the common trunk, there is always slight oxygen unsaturation of the arterial blood The degree of oxygen unsaturation of the arterial blood increases as the circulation

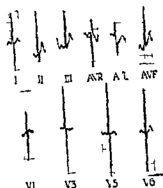


FIGURE XIV-21 Truncus arteriosus (same patient as in Figure XIV-9) Infant

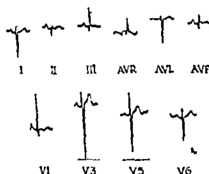


FIGURE XIV-22 Truncus arteriosus Child

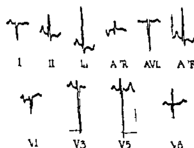


FIGURE XIV-23 Truncus arteriosus Adult

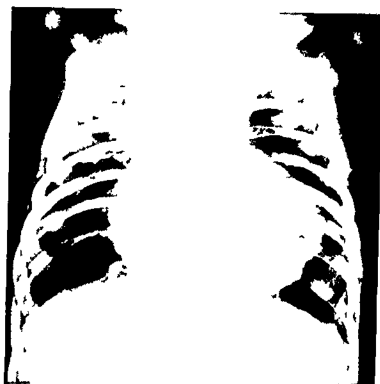
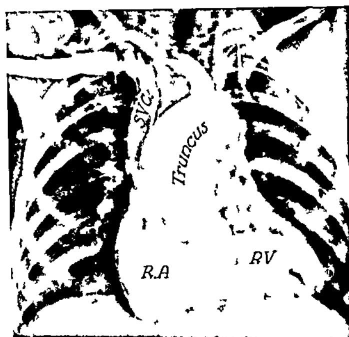


FIGURE XIV-20 Truncus arteriosus and a single ventricle with circulation to the lungs by way of the superior bronchial arteries (same patient as in Figure XIV-6) Infant

to the lungs is reduced When the pulmonary blood flow is meager, the oxygen saturation of the arterial blood may be extremely low Usually, however, the pulmonary blood flow is relatively constant and can be increased by exercise, therefore the oxygen saturation of the arterial blood drops but slightly, if at all, with exercise

The red blood cell count the amount of available hemoglobin and *the hematocrit reading* vary inversely with the adequacy of the pulmonary blood flow Inasmuch as there is always slight oxygen unsaturation of the arterial blood, even when the pulmonary blood flow is normal or excessive, there is always a slight increase in the red blood cell count, the amount of available hemoglobin, and the hematocrit reading Polycythemia increases as the oxygen unsaturation of the arterial blood increases The red blood cell count is frequently between 6 5 and 7 5 million cells per cu mm When the pulmonary blood flow is markedly reduced, the red blood cell count may reach 10 million and there is a proportional increase in the amount of available hemoglobin and in the hematocrit reading

Cardiac catheterization is of no great aid in diagnosis It is usually possible to catheterize the aorta The systolic pressure in the right ventricle is the same



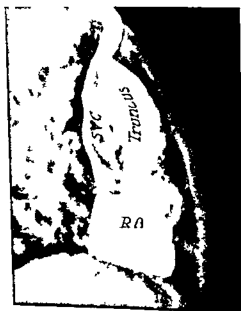


FIGURE XIV 24 Truncus arteriosus with reduced pulmonary blood flow Child

as that in the aorta, and the oxygen content of the blood in the aorta is usually higher than that in the right ventricle It is, however, impossible to catheterize the pulmonary artery directly from the right ventricle

Angiocardiography is of surprisingly little diagnostic aid When the pulmonary arteries arise directly from the truncus, the dye is dissipated so rapidly into the two circulations that frequently it cannot be traced beyond the right ventricle, and yet there is prompt disappearance of the dye from that chamber! When the circulation to the lungs is by way of the bronchial arteries, it is seldom possible to visualize these vessels accurately Angiocardiography is of no aid in the differentiation of this malformation from a tetralogy of Fallot with pulmonary atresia, because in neither instance can the dye enter the pulmonary artery in the normal manner In both instances dye must reach the lungs by way of the collateral circulation An abnormally large aorta," such as is shown in Figure XIV–24, is strongly suggestive of a truncus arteriosus The impression is confirmed by the delineation of a small vessel coursing to the lung

DIAGNOSIS

The occurrence of a continuous murmur over the lungs in a patient with persistent cyanosis and evidence of some cardiac abnormality is almost pathognomonic of a truncus arteriosus Therefore, in the presence of a continuous murmur, the diagnosis of a truncus arteriosus is relatively easy and can be made with a high degree of accuracy

The diagnosis is based upon a combination of findings There may be minimal cyanosis, no limitation of activity, and a loud continuous murmur, there may be moderately severe cyanosis considerable limitation of activity, and a continuous murmur localized over one particular area in the chest, or finally there may be intense cyanosis, marked clubbing and polycythemia, extreme limitation of exercise, and no murmur

The x ray findings are of real aid in diagnosis The heart is slightly to moderately enlarged and there is a concave curve at the base of the heart to the left of the sternum, the 'aorta is abnormally large and arches at an unusually high level, it may arch either to the right or to the left, in the hilar regions no left or right pulmonary artery is visible but numerous fine vascular markings radiate from the hilar regions, delineation of the esophagus with barium may or may not reveal anomalous retro-esophageal vessels The only exception occurs in the rare instance when both pulmonary arteries arise from the left side of the base of the common truncus and there is a conspicuous hilar dance, such patients have a continuous murmur and show little or no cyanosis

tions o cur in combination with another malformation which causes persistent cyanosis, the condition may closely simulate a truncus arteriosus Cardiac catheterization or angiocardiography may be necessary to differentiate the two conditions Either of the tests will show that the pulmonary artery arises from the right ventricle, thus excluding a truncus arteriosus

Rupture of an aneurysm from the sinus of Valsalva into the lesser circulation also causes a continuous murmur The murmur, however, usually develops abruptly in adult life furthermore, the murmur is best heard over the precordium, *not over the lungs* The condition usually leads to progressive cardiac enlargement and cardiac failure

An Eisenmenger complex with aortic insufficiency may be confused with a truncus arteriosus when the dia tolic element of the continuous murmur is maximal over the body of the heart The contour of the heart differentiates the two conditions In a truncus arteriosus the shadow at the base of the heart to the left of the sternum is concave, except in the rare instance in which the main pulmonary artery arises from the base of the heart and causes fullness of the pulmonary conus Under the latter circumstances cardiac catheterization may be necessary to differentiate the two malformations

A tetralogy of Fallot with persistent patency of the ductus arteriosus gives essentially the same findings The rarity of such a malformation is shown by the fact that in the author s experience every case in which a continuous murmur was recorded proved at autopsy to be a truncus arteriosus Indeed, among the first 1 000 patients who had a Blalock Taussig operation there was not a single instance of a patient over two years of age with a tetralogy of Fallot in whom the ductus arteriosus had remained patent Therefore, although it is possible to have a tetralogy of Fallot with persistent patency of the ductus arteriosus, the more probable diagnosis is a truncus arteriosus

A tetralogy of Fallot and pulmonary atresia after the obliteration of the ductus arteriosus may require differentiation from a pseudo truncus arteriosus In early infancy the contours of the heart in these two conditions are quite different In a tetralogy of Fallot with pulmonary atresia the heart is essentially normal in shape and is frequently phenomenally small In contrast to this, in a truncus arteriosus the heart is moderately enlarged and the contour is distinctive in that there is no fullness of the pulmonary conus and the ventricle extends abruptly out to the anterior chest wall as a sharp shelf Infants with a tetralogy of Fallot and pulmonary atresia almost always suffer from severe attacks of paroxysmal dyspnea whereas most infants with a truncus arteriosus, although extremely

DIFFERENTIAL DIAGNOSIS

When cyanosis is minimal *or absent*, a truncus arteriosus may require differentiation *from* a patent ductus arteriosus or an aortic septal defect, from peripheral pulmonary stenoses, from a rupture of an aneurysm of the sinus of Valsalva or even from an Eisenmenger complex with aortic insufficiency In the presence of moderate cyanosis a truncus arteriosus is usually mistaken for a tetralogy of Fallot with persistent patency of the ductus arteriosus In the presence of intense cyanosis, it may be mistaken for a tetralogy of Fallot with pulmonary atresia, for a non functioning right ventricle, or even for a transposition of the aorta combined with pulmonary atresia

Persistent patency of the ductus arteriosus differs from a truncus arteriosus in *that the oxygen saturation of the arterial blood* is normal The only exception *occurs in* early infancy, when the arterial oxygen saturation may be slightly reduced, under such circumstances it usually rises to normal with exercise (see Chapter XX) X ray and fluoroscopy show fullness of the pulmonary conus and conspicuous pulmonary arteries The electrocardiogram usually shows a balanced axis Cardiac catheterization clinches the diagnosis in that the pulmonary artery, not the aorta, is entered from the right ventricle

An aortic septal defect may be confused with a truncus arteriosus, especially when the pulmonary arteries arise from a common orifice on the posterior wall of the truncus arteriosus In a truncus arteriosus this orifice is the point of origin of the pulmonary circulation, whereas in an aortic septal defect the aorta and the pulmonary arteries arise normally from their respective ventricles and there *is merely a failure in the* completion of the aortic septum The differentiation of *these two* conditions is vitally important, as surgical closure of an aortic septal defect would restore the heart to normal, whereas closure of the orifice of the pulmonary artery in a truncus arteriosus would cut off all circulation to the lungs and kill the patient

In both conditions there may be a continuous murmur over the base of the heart Cyanosis may be minimal or absent The continuous murmur of an aortic septal defect more closely resembles that of a patent ductus arteriosus than that of a truncus arteriosus It is seldom as loud as that caused by a large ductus arteriosus In doubtful cases, cardiac catheterization may be necessary to determine whether the pulmonary artery or the aorta arises from the right ventricle

Peripheral pulmonary stenoses due to localized constrictions in the branches of the pulmonary arteries may cause a continuous murmur over the lungs which is closely similar to that produced by a truncus arteriosus When such constric

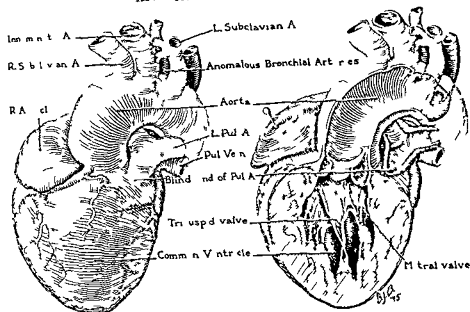


FIGURE XIV 25 Pseudo truncus arteriosus with a single ventricle

tigial pulmonary artery Inasmuch as all the blood returned to the heart is pumped out through a single great vessel, the occurrence of a single ventricle does not fundamentally alter the course of the circulation, as shown in Diagram xiv–3 The basic clinical findings are also unaltered by the absence of the ventricular septum Nevertheless, the occurrence of a single ventricle would be of extreme importance if total correction of the malformation were undertaken

TREATMENT

When the pulmonary arteries arise directly from the common trunk, the danger is that of excessive pulmonary blood flow and pulmonary hypertension In certain instances with excessive pulmonary blood flow and a conspicuous hilar dance, if it were possible to decrease the size of the pulmonary orifice and break the pressure to the lungs, such an operation would benefit the patient Fortunately, in the vast majority of patients the pulmonary vessels which arise from the aorta are smaller than normal and the condition is compatible with a long and active life

If the circulation to the lungs is by way of the small pathways of collateral circulation, the primary difficulty is the reduction of the pulmonary blood flow In such instances a Blalock Taussig operation may be of benefit In order to increase the pulmonary circulation it is not only essential to have a rudimentary

cyanotic at birth, do not suffer from paroxysmal dyspnea and generally do better than other infants with a corresponding degree of cyanosis If a patient with a *tetralogy of Fallot* and *pulmonary atresia* survives the closure of the ductus arteriosus, the condition is functionally that of a truncus arteriosus, except that the pulmonary vascular bed is normal because prior to the closure of the ductus arteriosus the pulmonary artery carried blood to the lungs

A non functioning right ventricle causes a square contour of the heart in the anterior posterior position Examination of the heart in the left anterior-oblique position differentiates the two conditions in a non functioning right ventricle, the cardiac shadow does not extend forward anterior to the aorta, whereas in a truncus arteriosus the right ventricle appears definitely enlarged Furthermore, in a non functioning right ventricle there is electrocardiographic evidence of left axis deviation and left ventricular hypertrophy

Complete transposition of the great vessels may occasionally be confused with a truncus arteriosus, because in both conditions there may be persistent cyanosis and relatively pronounced vascular markings Time soon differentiates the two conditions An infant with a complete transposition of the great vessels does poorly, the heart enlarges, and cyanosis increases An infant with a truncus arteriosus and increased flow, if he regains compensation, shows a marked decrease in cyanosis and usually develops a continuous murmur

Transposition of the aorta combined with pulmonary stenosis or atresia is associated with a large aorta, consequently a prominent aortic knob is visible in the anterior posterior position In the left anterior-oblique position the pulmonary window is abnormally clear, in the right anterior oblique position the cardiac shadow causes a sharp shelf like projection from the aorta toward the chest wall The vascular markings extend nearly to the periphery of the lungs when there *is* a transposition of the great vessels and pulmonary stenosis When the aorta is transposed so far to the left that it occupies the position of the normal pulmonary artery, the unusual course of the aorta and the clear lung fields aid in the differentiation of a transposition of the aorta from a truncus arteriosus (see Chapter x, Section c) The presence or absence of a continuous murmur is also of great diagnostic importance Occasionally the clinical differentiation between the two conditions is extremely difficult

COMMONLY ASSOCIATED MALFORMATIONS

A single ventricle may occur in combination with a truncus arteriosus Figure xiv-25 is a drawing of this type of malformation, in which there was a ves-

DIAGRAM XIV-3

Truncus arteriosus and a single ventricle

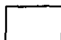
In this malformation there is but a single ventricle from which a single great vessel the truncus arteriosus arises The blood is pumped out through the truncus arteriosus to the body and by the pathways of the collateral circulation to the lungs The pulmonary artery if present is a blind tube which has no connection with the truncus arteriosus or with the common ventricle

The blood from the right auricle flows into the common ventricle and is pumped out through the truncus arteriosus to the systemic circulation The blood from the body is returned in the normal manner by the superior vena cava and the inferior vena cava to the right auricle and thence to the common ventricle Inasmuch as the pulmonary artery fails to meet the ventricle and the ductus arteriosus is absent the only way for the blood to reach the lungs is through the bronchial arteries or by some of the other pathways of the collateral circulation The blood which does reach the lungs is returned by the pulmonary veins to the left auricle thence the blood flows into the common ventricle There the cycle starts again

Clinical diagnosis Cyanosis is intense The heart is relatively normal in size but there is no fullness of the pulmonary conus During infancy the contour of the heart resembles that of a non functioning right ventricle As the child grows older the contour of the heart changes and becomes similar to that of a tetralogy of Fallot with an extremely severe pulmonary stenosis The large size of the aortic shadow which lies at an abnormally high level offers a clue to the correct diagnosis If the collateral circulation is by way of the superior bronchial arteries the arch of the aorta may appear fuzzy Unless the cyanosis is extremely intense and the lungs excessively clear, a continuous murmur is usually audible over some areas of the lungs The electrocardiogram generally shows a right axis deviation and evidence of combined hypertrophy

Diagram XIV-3

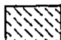
Head and upper extremities
Superior Bronchial Artery
R Lung
S.V.C.
PV
Truncus Arteriosus
L Lung
pul. V
L A
R A
Descending Aorta
Common Ventricle
I.V.C.
Trunk and lower extremities



Arterial blood (fully saturated)



Venous and arterial blood
Cyanosis visible



Small admixture of venous blood
No visible cyanosis



Venous blood

truncus arteriosus over rides the ventricular septum, a high ventricular septal defect *is an integral* part of the malformation Furthermore, since the truncus arteriosus receives blood from both ventricles, there is always some degree of oxygen unsaturation of the arterial blood

[illegible] adequacy of the pulmonary blood flow [illegible]

for exercise is moderately reduced Nevertheless, [illegible] with life for a number of years If, however, the circulation to the lungs is by way of small bronchial arteries or other minute vessels, cyanosis and polycythemia are intense, the patient s exercise tolerance is extremely limited

The heart is usually slightly to moderately enlarged, the second sound at the base is accentuated but never reduplicated The murmurs vary with the ade quacy of the pulmonary blood flow When the pulmonary blood flow is ade quate or excessive, there is a loud *continuous murmur, when it is moderately reduced, the continuous* murmur is usually localized to some specific portion of the chest when the pulmonary blood flow is minimal, there may be no murmur

X ray and fluoroscopy show slight to moderate cardiac enlargement, a large aorta, a high aortic arch, absence of fullness of the pulmonary conus, and absence of the normal shadows cast by the pulmonary arteries Usually the lung fields are clear and numerous fine shadows are seen to radiate from the hilar regions The esophagram may show evidence of retro-esophageal vessels

The electrocardiogram usually shows right axis deviation in the standard leads the precordial leads are variable but generally show evidence of hyper trophy of both ventricles

The circulation time is abnormally short

Cardiac catheterization and angiocardiography are of diagnostic aid mainly in the exclusion of other conditions

The diagnosis is based upon the finding of a continuous murmur over the lungs in a patient with persistent cyanosis, combined with characteristic x ray and fluoroscopic findings

In the absence of cyanosis a truncus arteriosus *may require* differentiation from a patent ductus arteriosus or an aortic septal defect, from *peripheral pul* monary stenoses from a rupture of an aneurysm of the sinus of Valsalva, or from an Eisenmenger complex with aortic insufficiency In the presence of cyanosis,

pulmonary artery of sufficient size to carry blood to the lungs, but also essential to have a normal pulmonary vascular bed If such is the case, since both auricles are normally formed and both ventricles pump the blood into the common trunk, the creation of an artificial ductus arteriosus increases the pulmonary blood flow without any significant alteration in the work required of the heart Under such circumstances a Blalock-Taussig operation is of great benefit

Unfortunately, in many instances of a pseudo truncus arteriosus, even though the pulmonary artery is of fair size, the pulmonary vascular bed appears unable to open up in the normal manner and carry the increased volume of blood to the lungs When such is the situation, thromboses develop distal to the anastomosis Not only does the patient fail to derive benefit from the operation but generally he does not survive The over all mortality rate for patients with truncus arteriosus is about 25 per cent Therefore, if the child is only slightly incapacitated, operation is contraindicated

PROGNOSIS

The prognosis depends upon the pulmonary blood flow If large pulmonary arteries arise directly from the common trunk, the infant may die of excessive pulmonary blood flow within the first few days

If the pulmonary artery arises from the truncus arteriosus at a slightly higher level or from a common orifice, the condition may be compatible with relative longevity

Indeed, if the collateral circulation is such that there is a relatively adequate pulmonary blood flow, the prognosis is reasonably good Even though the patient is cyanotic, there is only moderate polycythemia, his exercise tolerance will be good and for many years he may enjoy a relatively normal life

If the pulmonary blood flow is markedly reduced, a Blalock Taussig operation is indicated If this is successful, the prognosis will be greatly improved Without operation, the prognosis is poor

SUMMARY

A truncus arteriosus is the result of an early arrest in the development of the great vessels, either the pulmonary artery arises directly from the base of the common trunk, or a vestigial pulmonary artery fails to reach the aortic trunk and the circulation to the lungs is by way of the bronchial arteries In both instances the blood from the two ventricles is pumped out into a common vessel which directs the blood to the body and to the lungs Both ventricles pump against systemic pressure, both ventricles are thick walled Inasmuch as the

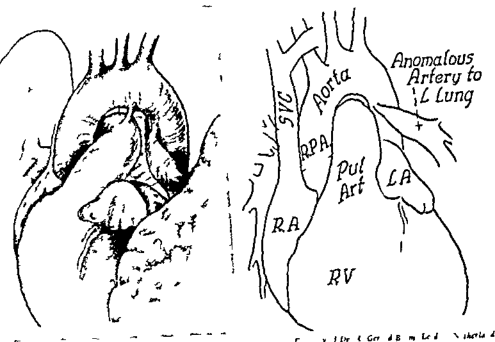


FIGURE XIV–26 Hemi truncus arteriosus (same patient as in Figures XIV–28 29) Child

COURSE OF THE CIRCULATION

During fetal life although the single pulmonary artery goes to only one lung, so little blood flows through the pulmonary artery that the absence of the other pulmonary artery places no strain on the fetal circulation At birth the heart is normal in size (see Figure XIV–27)

After birth the blood from the right auricle flows into the right ventricle and thence is pumped out into the main pulmonary artery and through the single pulmonary vessel to one lung, where it is oxygenated, and returned in the normal manner to the left auricle The blood from the left auricle flows into the left ventricle and is pumped out through the aorta to the systemic circulation and also to the lung which receives its blood from the anomalous vessel that arises from the aorta This lung receives fully oxygenated blood, circulates through the lung, and is then returned to the left auricle The blood which circulates through the body is returned in the normal manner by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again (see Diagram XIV–4)

it is frequently mistaken for a tetralogy of Fallot with persistent patency of the ductus arteriosus When cyanosis is intense, a truncus arteriosus is anatomically and functionally closely related to that which occurs in a patient with a tetralogy of Fallot combined with pulmonary atresia who survives the closure of the ductus arteriosus Nevertheless, the difference between these two conditions is important because of the difference in the operative risk Occasionally the condition requires differentiation from a complete transposition of the great vessels with or without pulmonary atresia or from a non functioning right ventricle

A patient with severe reduction in the pulmonary blood flow may be greatly benefited by a Blalock-Taussig operation, provided not only that a vestigial pulmonary artery exists, but also that the pulmonary vascular bed is normal and the pulmonary artery is able to direct blood to the lungs in the normal manner The mortality rate from operation is approximately 25 per cent

The prognosis varies with the adequacy of the pulmonary blood flow If the pulmonary blood flow is relatively adequate, the prognosis is excellent When the pulmonary blood flow is severely reduced, the prognosis may be greatly improved by successful surgery

B *Hemi Truncus Arteriosus*

In a hemi truncus arteriosus, one pulmonary artery arises directly from the aorta, as it does in a truncus arteriosus, and the other pulmonary artery arises normally from the right ventricle Although a single pulmonary artery occurring in combination with various malformations, such as a tetralogy of Fallot, may be considered as a variant of a hemi truncus arteriosus, the following discussion is concerned with the condition when it occurs as an isolated abnormality

NATURE OF THE MALFORMATION

"Hemi truncus arteriosus" is the name given to the condition in which the heart itself is normal and the great vessels arise normally but the main pulmonary artery supplies only one lung and the other lung is supplied by a vessel which arises directly from the aorta The single pulmonary artery may go to either the right or the left lung The author has studied one case in which the pulmonary artery went to the left lung and Dr A Gerard Brom told her of a case in which the pulmonary artery was directed to the right lung (see Figure XIV-26) In both instances the circulation to the other lung was by way of an anomalous vessel which arose from the aorta

CARDIAC FINDINGS

... The *heart sounds* are normal There is,

aorta When the murmur is heard on the left side, ...
of persistent patency of the ductus arteriosus The murmur has a more humming quality and is usually more widely transmitted than that of a patent ductus arteriosus

X RAY FINDINGS

The x ray findings are not remarkable (see Figure XIV–28)

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram is usually normal

SPECIAL TESTS

Cardiac catheterization shows that the circulation is normal Indeed, when patency of the ductus arteriosus is suspected, the absence of any increase in the oxygen content of the blood in the pulmonary artery gives the clue to the diagnosis of a hemi truncus arteriosus

Angiocardiography is diagnostic in that one pulmonary artery is visualized with the right side of the heart and the other with the left The pulmonary artery which arises from the right ventricle fills normally (see Figure XIV–29) but no dye enters the other lung until the aorta is visualized and then only if the vessel is sufficiently large to permit opacification of the pulmonary vascular bed

DIAGNOSIS

A hemi truncus arteriosus may be suspected when a continuous murmur is well heard over the right lung, especially in an asymptomatic patient with a normal left aortic arch When the continuous murmur is heard over the left side of the chest it may easily be confused with that of patency of the ductus arteriosus

DIFFERENTIAL DIAGNOSIS

The condition may closely simulate patency of the ductus arteriosus, an aortic septal defect or peripheral stenoses of the pulmonary artery All these conditions are readily differentiated by angiocardiography or by cardiac catheterization A hemi truncus arteriosus, however, is so rare that neither of these

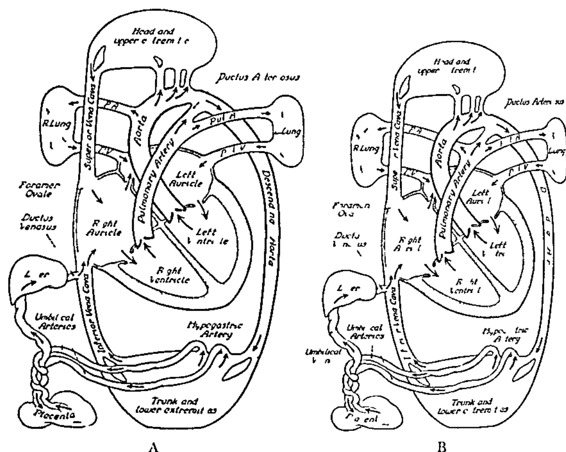


FIGURE XIX-27 Fetal circulation (A) Hemi truncus arteriosus and (B) normal heart

PHYSIOLOGY OF THE MALFORMATION

The entire oxygenation of the blood occurs in the lung which receives venous blood from the pulmonary artery The lung which receives blood from the aorta is non functioning as far as oxygenation is concerned, but does aid in the normal expansion of the chest and prevents the displacement of the mediastinum The outstanding physiological change caused by this anomaly is the difference in the amount of oxygen taken up and the amount of carbon dioxide given off in the two lungs Aside from this abnormality, the basic hemodynamics are normal

CLINICAL FINDINGS

The patient is generally asymptomatic The condition is usually detected on a routine physical examination

Cyanosis is absent There is no admixture of venous and arterial blood, the oxygen saturation of the arterial blood is normal

DIAGRAM XIV-4

Hemi truncus arteriosus

The essential feature of this malformation is that there is but a single pulmonary artery which goes to only one lung the other lung receives its blood from a vessel which arises from the aorta

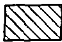
The blood from the right auricle flows into the right ventricle and is pumped out into the pulmonary artery, thence it flows to one lung only In that lung the blood is oxygenated in the normal manner and is returned to the left auricle The blood from the left auricle flows into the left ventricle and is pumped out through the aorta to the systemic circulation and through an anomalous artery (A A) to the other lung The fully oxygenated blood thus directed to the lungs through the anomalous vessel passes through the capillaries and is returned to the left auricle It follows that all the blood which the left auricle receives is fully oxygenated this blood flows into the left ventricle and out into the aorta The major part of the blood in the aorta is directed to the systemic circulation and the venous blood from the body is returned by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again

Clinical diagnosis The patient is asymptomatic and shows no cyanosis The heart is normal in size and shape A continuous murmur is audible over the lung on the side which has the anomalous circulation The electrocardiogram is normal

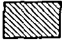
Diagram XIV-4

Head and upper extremities
R Lung
A.A.
Aorta
S.V.C.
P.V.
Pul. A.
L Lung
Pul. V.
L A
Pulmonary A
R.A.
L V
R.V.
Descending Aorta
I.V.C.
Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

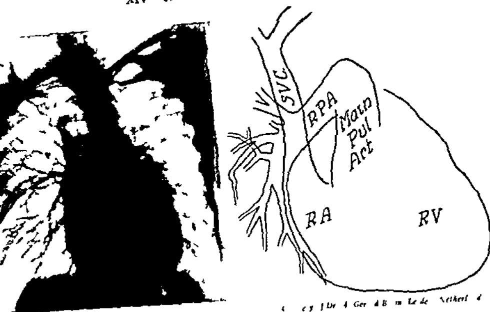


FIGURE XII-29 Hemi truncus arteriosus (same patient as in Figure XII-26) Child

Chapter XXI) Although the patient is living on one lung and the other lung is of no use in the oxygenation of the blood, there is no evidence that the functionless lung is prone to infection Actually that lung aids in the normal growth and expansion of the chest Hence pneumonectomy is contraindicated

SUMMARY

A hemi truncus arteriosus means that the circulation to one lung is similar to that of a truncus arteriosus and the circulation to the other lung is normal Thus a single pulmonary artery supplies one lung and an anomalous vessel which arises from the aorta directs the blood to the other lung Although the entire oxygenation of the blood occurs in the lung which has a normal supply, the patient is asymptomatic The existence of an abnormality is readily detected upon physical examination, as there is a continuous murmur over the lung which receives its blood from the aorta When this murmur is heard on the left side, the condition may be mistaken for patency of the ductus arteriosus

The condition requires differentiation from persistent patency of the ductus arteriosus and from peripheral pulmonary stenoses Both these conditions can readily be differentiated from a hemi truncus arteriosus by angiocardiography When there is but a single pulmonary artery the dye passes from the right ven



C[illegible]y of Dr [illegible] Ger[illegible]d B [illegible] Le[illegible]de[illegible] the [illegible]

FIGURE XIV-28 Hemi truncus arteriosus (same patient as in Figure XIV-26) Child

procedures is indicated as a routine to exclude the possibility in a patient with classic signs of patency of the ductus arteriosus An occasional exploratory thoracotomy carries no greater risk than would a routine angiocardiogram performed on every patient suspected of patency of the ductus arteriosus

Peripheral pulmonary stenoses are usually associated with hypertension in the proximal portion of the pulmonary artery This causes accentuation of the second sound at the base of the heart to the left of the sternum and, furthermore, the electrocardiogram usually shows a right axis deviation and evidence of right ventricular hypertrophy

TREATMENT

There is none, and none is necessary If operation is performed for patency of the ductus arteriosus and none is found, the chest should be closed unless further dissection is deemed advisable to rule out an aortic septal defect (see

CHAPTER XV

A SINGLE VENTRICLE AND A RUDIMENTARY OUTLET CHAMBER

A SINGLE ventricle is the result of an extremely early arrest in the develop ment of the heart It produces an entirely different architecture of the ventricles from that found in any other malformation [1]

Furthermore, the failure of the formation of the interventricular septum is frequently associated with variations in the size and position of the great vessels Either great vessel may be large, either may be small In addition there may be a transposition of the great vessels Consequently there is great variation in the clinical picture The diagnosis is correspondingly difficult and is frequently made only by the exclusion of other conditions Nevertheless, it is an important malformation

A single ventricle is relatively common when the primitive cardiac loop swings to the right instead of developing in the normal manner with the apex of the heart pointing to the left Hence it is common in patients with dextrorota tion of the heart A single ventricle, however, may occur when the heart occupies its normal position

A brief review of the embryology will help to clarify its nature.

EMBRYOLOGY

When the primitive cardiac tube bulges forward and swings to the right, the anterior portion of it becomes the bulbus cordis and the posterior part of the loop forms the common ventricle A single great vessel arises from the bulbus *cordis* Normally, as the heart develops, the ridge which separates the bulbus cordis from the *common ventricle atrophies* Coincidentally with the disappearance of this ridge the aortic septum develops and divides the aortic trunk into the aorta and the pulmonary artery At the same time the development of the ventricular septum divides the common ventricle into two chambers The posterior portion of the common ventricle becomes the left ventricle, from which the aorta arises, the lower portion of the bulbus cordis expands and fuses with the anterior por tion of the common ventricle to form the right ventricle, from which the pulmo- nary artery arises

In the malformation under discussion the ventricular *septum* fails to develop

tricle to one lung only, and no dye reaches the other lung until after the aorta has been visualized

A hemi truncus arteriosus causes no difficulty and requires no treatment In deed, pneumonectomy is contraindicated

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casionally both vessels may arise from the diminutive chamber, more often there is a transposition of the great vessels Under the latter circumstances the aorta arises from the diminutive chamber and the pulmonary artery from the common ventricle Ordinarily the vessel which arises from the rudimentary chamber is diminutive in size and that which arises from the common ventricle is of normal size or enlarged It is, however, theoretically possible for the pulmonary artery to be stenotic when it arises from the common ventricle

The malformation is further complicated when a transposition of the great vessels is combined with the anomalous insertion of the chordae tendineae of the tricuspid valve along the base of the rudimentary outlet chamber in such a manner that the blood from the right auricle is directed primarily into the rudimentary chamber and hence to the transposed aorta Figure xv-2 shows a drawing of such a specimen and Figure xv-3 shows the manner in which the blood is directed from the two auricles into the common ventricle and out into the aorta and the pulmonary artery This combination of anomalies, which was reported by Lambert is termed the Lambert heart The malformation is not rare, it produces a distinctive clinical picture

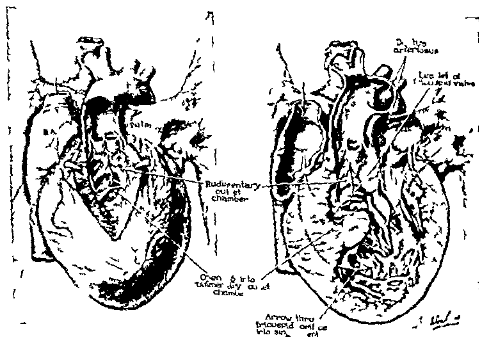


FIGURE XV 2 Single ventricle (Lambert heart) (same patient as in Figures XV 3 8,9) Child

and the bulbus cordis persists as a rudimentary outlet chamber. The arrest in the development of the heart may occur so early that both great vessels arise from the persistent bulbus cordis, or one may arise from the common ventricle and one from the rudimentary outlet chamber.

NATURE OF THE MALFORMATION

The essential feature of the malformation is a single ventricle with a rudimentary outlet chamber which lies in the region normally occupied by the outflow tract of the right ventricle. The diminutive chamber is separated from the main ventricle by a muscular ridge (see Figure xv-1). No valve or membrane guards its orifice. There is free communication between the common ventricle, which receives the blood from both auricles, and the small chamber from which one or both the great vessels arise. If the great vessels develop normally, the aorta arises from the common ventricle and the pulmonary artery from the rudimentary chamber. It is, however, common to find that the arrest in the development of the ventricle is associated with an abnormality of the great vessels. Oc-

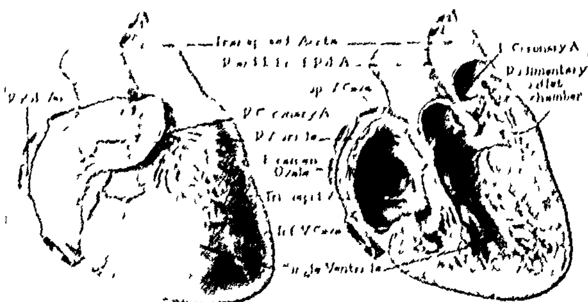


Figure xv-1. Single ventricle and a rudimentary outlet chamber from which the transposed aorta arises (same patient as in Figure x-7). Infant.

The foramen ovale is incompletely covered by a valve. In this case the mitral valve was atretic. Usually it, too, opens into the common ventricle.

casionally both vessels may arise from the diminutive chamber, more often there is a transposition of the great vessels *Under the latter circumstances the* aorta arises from the diminutive chamber and the pulmonary artery from the com mon ventricle *Ordinarily the vessel which arises from the rudimentary chamber* is diminutive in size and that which arises from the common ventricle is of nor *mal size or enlarged It is, however, theoretically possible for the* pulmonary artery to be stenotic when it arises from the common ventricle

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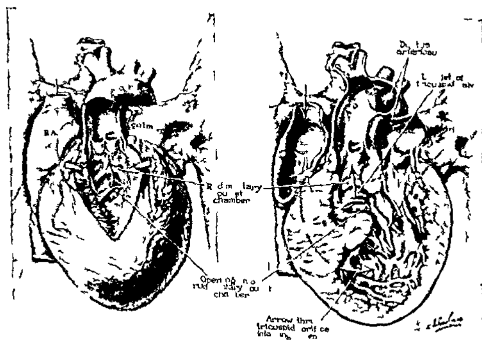


Figure xv 2 Single ventricle (Lambert heart) (same patient as in Figures xv 3 8 9) Child

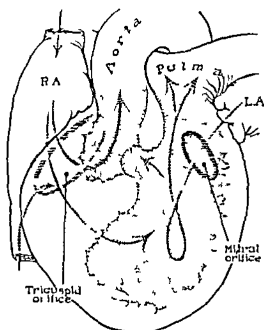


FIGURE xi–3 Single ventricle (Lambert heart) (same patient as in Figure xi–2) Child

The arrest in the development of the ventricles may or may not be associated with a malformation of the auricular septum If the auricular septum develops normally, the heart is a triloculate one or *cor triloculare biatriatum* (three-chambered heart with one ventricle and two auricles) Under such circumstances the mitral and tricuspid valves open into the common ventricle If, on the other hand, there is an arrest in the development of the auricular septum so that it, too, fails to form, the resultant malformation is a *cor biloculare,* or a biloculate heart *with a single auricle, a common atrioventricular valve, and a single ventricle*

COURSE OF THE CIRCULATION

During fetal life, although the structure of the heart is extremely primitive, it suffices to meet the needs of the fetus Inasmuch as the lungs are of no functional importance, it makes little difference which of the great vessels is given off from the diminutive chamber and which from the common ventricle At birth the heart is normal in size

After birth the fact that one of the great vessels is diminutive in size has a profound effect upon the circulation The blood from the right auricle flows through *the tricuspid valve into the common ventricle and the blood from the* left auricle flows through the mitral valve into the common ventricle Therefore, regardless of the structure of the auricular septum, there is complete admixture of the venous and arterial blood in the common ventricle As the ventricle fills, blood also flows into the rudimentary chamber Consequently the same admix

ture of venous and arterial blood is pumped out into both great vessels If both great vessels arise from the diminutive chamber and are approximately the same size, approximately the same volume of blood is pumped out through the pul monary artery to the lungs as is pumped out through the aorta to the systemic circulation The blood from the lungs is returned by the pulmonary veins to the left auricle and the blood from the body is returned by the superior and inferior venae cavae to the right auricle The blood from both auricles again flows into the common ventricle Inasmuch as the pulmonary blood flow is approximately equal to the systemic blood flow, there may be no visible cyanosis Frequently, however, a relatively small volume of blood reaches the lungs for oxygenation and the systemic circulation is also meager, therefore the infant shows persistent cyanosis (see Diagram xv–1)

As a rule, one great vessel is given off the main ventricle and the other off the rudimentary chamber, and furthermore the vessel which is given off the main chamber is usually the larger of the two Consequently, although the course of the circulation is fundamentally the same, owing to the difference in the size of the two vessels, the resultant admixture of venous and arterial blood is quite different

When the pulmonary artery arises from the main ventricle and the aorta from the rudimentary chamber, a large volume of blood is directed to the lungs and a relatively small amount is directed to the body Hence a large amount of fully oxygenated blood is returned to the left auricle, whereas a relatively small amount of venous blood is returned to the right auricle Consequently a large volume of oxygenated blood is mixed with a small amount of venous blood and the resultant admixture is above the threshold of visible cyanosis (see Diagram xv–2) Cyanosis is absent

When the aorta arises from the common ventricle and the pulmonary artery from the diminutive chamber, the situation is quite different Under such cir cumstances the pulmonary artery may be diminutive or the pulmonary orifice may be stenotic Thus the body receives an adequate or excessive volume of blood but only a small volume of blood goes to the lungs Hence a small volume of oxygenated blood is returned to the left auricle to mix with a large volume of venous blood returned from the body by the superior and inferior venae cavae to the right auricle Hence cyanosis is intense (see Diagram xv–3)

When a transposition of the great vessels is combined with the anomalous in sertion of the chordae tendineae of the tricuspid valve into the base of the rudi mentary chamber the situation is still further complicated Under such circum

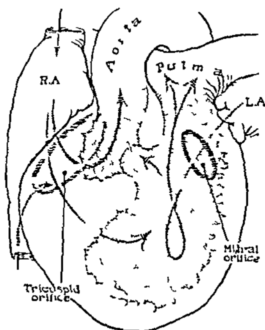


FIGURE xi-3 Single ventricle (Lambert heart) (same patient as in Figure xi-2) Child

The arrest in the development of the ventricles may or may not be associated with a malformation of the auricular septum If the auricular septum develops normally, the heart is a triloculate one or *cor triloculare biatriatum* (three-chambered heart with one ventricle and two auricles) Under such circumstances the mitral and tricuspid valves open into the common ventricle If, on the other hand, there is an arrest in the development of the auricular septum so that it, too, fails to form, the resultant malformation is a *cor biloculare* or a biloculate heart with a single auricle, a common atrioventricular valve, and a single ventricle

COURSE OF THE CIRCULATION

During fetal life, although the structure of the heart is extremely primitive, it suffices to meet the needs of the fetus Inasmuch as the lungs are of no functional importance, it makes little difference which of the great vessels is given off from the diminutive chamber and which from the common ventricle At birth the heart is normal in size

After birth the fact that one of the great vessels is diminutive in size has a profound effect upon the circulation The blood from the right auricle flows through the tricuspid valve into the common ventricle and the blood from the left auricle flows through the mitral valve into the common ventricle Therefore, regardless of the structure of the auricular septum, there is complete admixture of the venous and arterial blood in the common ventricle As the ventricle fills, blood also flows into the rudimentary chamber Consequently the same admix

DIAGRAM XV–I

Single auricle and a single ventricle with a rudimentary outlet chamber from which both great vessels arise

In this malformation the auricular septum is so rudimentary that there is but a single large auricle Both the tricuspid valve and the mitral valve may be present or there may be a common atrioventricular valve which opens into the ventricle There is but a single ventricle with a rudimentary outlet chamber from which both the pulmonary artery and the aorta arise The great vessels are diminutive in size

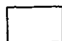
The blood from the common auricle flows into the common ventricle Inasmuch as both great vessels arise from the rudimentary outlet chamber, all the blood from the common ventricle must be forced from this small chamber into the aorta and the pulmonary artery Since both great vessels arise from the same chamber, the pressure in the aorta and the pulmonary artery is the same The blood which is pumped into the aorta goes to the systemic circulation and is returned by the superior vena cava and the inferior vena cava to the right side of the auricle. The blood which is pumped into the pulmonary artery goes to the lungs where it is oxygenated, and is returned by the pulmonary veins to the left side of the auricle There the cycle starts again

Clinical diagnosis is based upon the contour of the heart The size of the heart is not greatly altered but the rudimentary outlet chamber causes a prominence of the pulmonary conus in the anterior posterior position in the left anterior-oblique position however there is no enlargement of the right ventricle The increased pressure in the ventricle is transmitted to the auricle and this, in turn, causes dilatation of the superior vena cava

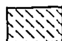
There is complete admixture of the venous and arterial blood in both the common auricle and the single ventricle Furthermore inasmuch as the two great vessels are of equal size approximately equal volumes of blood are directed to the pulmonary and systemic circulations there is persistent cyanosis The mechanism for pumping the blood from the single ventricle through the rudimentary outlet chamber is inefficient The condition is rarely compatible with life for more than a few months

DIAGRAM XV-1

Head and upper extremities
R Lung
S.V.C.
P.A.
Aorta
Pul A
L Lung
PV
Pul V
Pulmonary A
L A
R.A.
Descending Aorta
Rudimentary Outlet Chamber
Single Ventricle
I.V.C.
Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

Diagram XV-1

Single auricle and a single ventricle with a rudimentary outlet chamber from which both great vessels arise

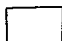
In this malformation the auricular septum is so rudimentary that there is but a single large auricle Both the tricuspid valve and the mitral valve may be present or there may be a common atrioventricular valve which opens into the ventricle. There is but a single ventricle with a rudimentary outlet chamber from which both the pulmonary artery and the aorta arise The great vessels are diminutive in size

The blood from the common auricle flows into the common ventricle Inasmuch as both great vessels arise from the rudimentary outlet chamber, all the blood from the common ventricle must be forced from this small chamber into the aorta and the pulmonary artery Since both great vessels arise from the same chamber the pressure in the aorta and the pulmonary artery is the same The blood which is pumped into the aorta goes to the systemic circulation and is returned by the superior vena cava and the inferior vena cava to the right side of the auricle The blood which is pumped into the pulmonary artery goes to the lungs where it is oxygenated and is returned by the pulmonary veins to the left side of the auricle. There the cycle starts again

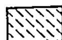
Clinical diagnosis is based upon the contour of the heart The size of the heart is not greatly altered but the rudimentary outlet chamber causes a prominence of the pulmonary conus in the anterior posterior position in the left anterior-oblique position however there is no enlargement of the right ventricle The increased pressure in the ventricle is transmitted to the auricle and this in turn causes dilatation of the superior vena cava

There is complete admixture of the venous and arterial blood in both the common auricle and the single ventricle. Furthermore, inasmuch as the two great vessels are of equal size approximately equal volumes of blood are directed to the pulmonary and systemic circulations there is *persistent cyanosis* The mechanism for pumping the blood from the single ventricle through the rudimentary outlet chamber is inefficient The condition is rarely compatible with life for more than a few months

Diagram xv-2


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

Diagram XV–2

Single ventricle with a rudimentary outlet chamber from which the aorta arises

In this malformation there are two auricles and a single ventricle with a rudimentary outlet chamber The great vessels are transposed The pulmonary artery arises from the main ventricle and is of normal size the aorta arises from the diminutive outlet chamber and is of small caliber

The blood from both auricles flows into the common ventricle As the common ventricle fills the blood flows into the rudimentary outlet chamber The blood from the common ventricle is pumped into the pulmonary artery and that from the rudimentary chamber into the aorta The blood in the pulmonary artery flows to the lungs and the oxygenated blood is returned by the pulmonary veins to the left auricle The blood in the aorta flows into the systemic circulation and is returned in the normal fashion by the superior and inferior venae cavae to the right auricle Regardless of whether or not the auricular septum is sufficiently well developed to prevent an appreciable admixture of the blood in the two auricles, there is complete admixture of venous and arterial blood in the common ventricle. Inasmuch as the pulmonary artery is larger than the aorta and is given off the common ventricle, a far greater volume of blood flows to the lungs than to the systemic circulation Furthermore, the blood is pump d to the lungs under systemic pressure, hence there is severe pulmonary hypertension Nevertheless the volume of oxygenated blood returned to the left auricle is larger than the volume of venous blood returned to the right auricle. Usually the volume of oxygenated blood returned from the lungs is sufficiently great so that there is no visible cyanosis

Clinical diagnosis is based upon the contour of the heart In infancy, in the anterior posterior position the rudimentary outlet chamber causes a prominence in the region of the pulmonary conus In older children this chamber is no longer visible and the aortic shadow is narrow In both instances, in the left anterior-oblique position, there is no enlargement of the right ventricle. In addition, there is usually great exaggeration of the hilar shadows due to the large volume of blood flowing through the pulmonary circulation Clubbing and cyanosis are absent Murmurs may or may not be present and are of no diagnostic aid The electrocardiogram is variable but may offer a clue to the diagnosis in that there is often a discrepancy between the axis deviation in the standard leads and the evidence of ventricular hypertrophy in the unipolar precordial leads

Diagram XV-3

Head and upper extremities
R Lung
L Lung
S.V.C.
P.A.
Aorta
Pul. A.
PV
Pul V
Pulmonary Art.
L.A.
R.A.
Single Ventricle
Rudimentary Outlet Chamber
Descending Aorta
I.V.C.
Trunk and lower extremities

Arterial blood (fully saturated)

Venous and arterial blood. Cyanosis visible

Small admixture of venous blood. No visible cyanosis

Venous blood

DIAGRAM XV-3

Single ventricle with a rudimentary outlet chamber from which the pulmonary artery arises

This malformation represents an early arrest in the development of the heart There is but a single ventricle with a rudimentary outlet chamber The aorta lies more posteriorly than does the pulmonary artery It is given off the common ventricle and is of normal size, whereas the pulmonary artery is given off the *rudimentary chamber* and is of small caliber

The blood from the right auricle together with that from the left auricle passes [illegible] as the common ventricle [illegible] The blood in the common ventricle is pumped into the aorta and that in the rudimentary outlet chamber is pumped into the pulmonary artery The blood in the aorta flows into the systemic circulation and is returned in the normal fashion by the superior and inferior venae cavae [illegible]

Inasmuch as the aorta is large and arises from the common ventricle and the pulmonary artery is small and receives only the blood from the rudimentary chamber a *much greater volume of blood goes to the systemic circulation than goes to the lungs* for oxygenation Hence the volume of venous blood returned to the right auricle is larger than the volume of oxygenated blood returned to the left auricle Thus in the *common* ventricle there is admixture of a large volume of venous blood with a small *volume of oxygenated blood* Cyanosis is intense.

Clinical diagnosis is based upon the finding of persistent cyanosis and a heart with a contour which is essentially the same as that in other cases of a single ventricle with a rudimentary outlet chamber In infancy the heart is not enlarged nevertheless in the anterior posterior position there is prominence of the pulmonary conus, but in the left anterior-oblique position there is no enlargement of the right ventricle In older children the prominence of the pulmonary conus disappears and the contour of the heart resembles that of an extreme tetralogy of Fallot Cyanosis is intense The condition is usually compatible with life for only a few months

stances the blood from the right auricle is directed mainly to the rudimentary outlet chamber and thence directed into the transposed aorta The blood from the left auricle is directed into the common ventricle Consequently even though the pulmonary artery may be greatly enlarged, the systemic circulation receives mainly venous blood The patient will be intensely cyanotic The course of the circulation is shown in Diagram xv-4

PHYSIOLOGY OF THE MALFORMATION

The two features of the common ventricle which affect the physiology of the malformation are the free admixture of venous and arterial blood in the common ventricle and the common ejectile force with which the blood from the ventricle is pumped into both great vessels Thus there is always some oxygen unsaturation of the arterial blood and, except when there is pulmonary stenosis, there is always pulmonary hypertension The high pressure with which the blood is ejected into the lungs eventually leads to intimal proliferation which causes progressive narrowing of the pulmonary vascular bed and thereby increases the pulmonary resistance Thus over a period of years less blood is pumped through the pulmonary circulation and more blood is directed into the aorta This means that the volume of blood which reaches the lungs for oxygenation is decreased, hence a smaller volume of oxygenated blood is returned to the left auricle and the oxygen content of the blood in the common ventricle is proportionally reduced Consequently, although a progressively greater volume of blood is directed to the systemic circulation, the oxygen saturation of the arterial blood is gradually reduced As the oxygen unsaturation slowly increases, cyanosis, even if not initially obvious, sooner or later becomes apparent and gradually deepens These changes, however, take years to develop Needless to say, in the presence of pulmonary stenosis, pulmonary hypertension never develops

CLINICAL FINDINGS

Inasmuch as a single ventricle may be associated with decreased or increased pulmonary blood flow, and furthermore the increased pulmonary blood flow may be associated with an anomalous insertion of the tricuspid valve of such a nature as to direct venous blood to the aorta, the clinical manifestations are protean

The history is often significant Infants who show no cyanosis frequently suffer from bronchitis and pneumonia Moreover, owing to the rapidity of the respirations and the congestion in the lungs, the condition is frequently misdiagnosed as pneumonia

If there is severe pulmonary stenosis or functional pulmonary atresia, the infant will suffer from severe *attacks of paroxysmal dyspnea* as the ductus arteriosus undergoes obliteration When the pulmonary artery is of small caliber or the pulmonary orifice only slightly stenotic, although cyanosis is *intense*, the infant does better than would be expected from the intensity of the cyanosis

Growth and development are usually retarded

The appearance of the patient may suggest the existence of a cardiac abnormality The child who shows no cyanosis frequently has a *gracile habitus* because of the large pulmonary blood flow and the small and rapid systemic flow Consequently, even though the oxygen saturation of the arterial blood is nearly normal, the systemic circulation is starved

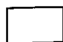
The presence or absence of cyanosis depends upon the volume of blood which reaches the lungs for oxygenation and the volume of venous blood which is directed into the aorta This, in turn, depends upon the size and position of the pulmonary artery and the relation of the tricuspid valve to the rudimentary outlet chamber When the pulmonary artery arises from the rudimentary outlet chamber it is usually of small caliber and may be stenotic or even atretic at its base Under such circumstances only a small volume of blood reaches the lungs for oxygenation and a small volume of oxygenated blood is mixed with a large volume of venous blood Cyanosis is intense

If, however, there is a transposition of the great vessels, so that the pulmonary artery is given off the main ventricle, it is usually of normal caliber, and the aorta, which arises from the rudimentary chamber, is of small caliber Under such circumstances a greater volume of blood goes to the lungs for oxygenation than to the systemic circulation Hence a large volume of oxygenated blood is mixed with a relatively small volume of venous blood There is usually *no* visible cyanosis

When there is a Lambert heart, that is, when a complete transposition of the great vessels is combined with an anomalous insertion of the tricuspid valve of such a nature that the major part of the venous blood from the right auricle is directed into the rudimentary outlet chamber and thence into the aorta, even though the pulmonary blood flow is large, the patient will show intense cyanosis

In brief, cyanosis is always present if there is decreased pulmonary blood flow but may be present when the pulmonary blood flow is excessive *Under the* latter circumstances the pulmonary artery arises posteriorly from the main ventricle and the occurrence of cyanosis is due to the anomalous insertion of the tricuspid valve which directs the blood from the right auricle into the rudimentary outlet chamber

DIAGRAM XV–4


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

DIAGRAM XV–4

Single ventricle with a rudimentary outlet chamber from which the transposed aorta arises (Lambert heart)

In this malformation there are two auricles and a single ventricle with a rudimentary outlet chamber and the great vessels are transposed There may or may not be an auricular septal defect The mitral valve opens into the single ventricle and the chordae tendineae of the tricuspid valve are attached to the wall of the outlet chamber

Owing to the abnormality of the tricuspid valve most of the blood from the right auricle is directed into the rudimentary outlet chamber and thence is pumped out through the aorta to the systemic circulation and is again returned by the superior vena cava and the inferior vena cava to the right auricle The blood in the left auricle flows through the mitral valve into the common ventricle A large volume of the blood in the common ventricle is pumped out through the pulmonary artery to the lungs, where it is oxygenated and returned by the pulmonary veins to the left auricle thence it again flows into the common ventricle Nevertheless since there is a common ventricle some of the blood which flows through the tricuspid valve flows into the common ventricle and some of the blood in the common ventricle is pumped into the rudimentary chamber and thence out into the aorta Therefore although there is a transposition of the great vessels and a large proportion of the venous blood is pumped out into the aorta there is considerable mixing of venous and arterial blood in the common ventricle The origin of the pulmonary artery from the common ventricle causes severe pulmonary hypertension

Clinical diagnosis Cyanosis is intense and dates from birth Nevertheless there is some constant crossing of the two circulations in the common ventricle hence the patient is not as limited as might be expected from the intensity of the cyanosis Furthermore the condition is relatively stable and compatible with life for a number of years The child does not squat when tired The heart is slightly enlarged but there is no progressive cardiac enlargement There is absence of fullness of the pulmonary conu but the pulmonary vascularity is markedly increased and there may be a hilar dance The electrocardiogram frequently shows a discrepancy between the axis deviation in the standard leads and the evidence of ventricular hypertrophy in the unipolar precordial leads Pulmonary hypertension increases with age and usually causes progressive incapacity in early adult life

Clubbing of the extremities is directly proportional to the cyanosis If the patient shows no visible cyanosis and the red blood cell count is but slightly elevated, clubbing is absent If cyanosis and polycythemia are marked, clubbing develops at an early age

The exercise tolerance of the individual varies with the oxygen saturation of the arterial blood Nevertheless, since the patient is able to increase the pulmonary blood flow with exercise, his tolerance is better than is expected and he does not squat when tired For this reason, the possibility of a single ventricle should be considered when a child who is intensely cyanotic has a fair exercise tolerance Nevertheless, over a period of years the patient gradually becomes progressively more incapacitated

CARDIAC FINDINGS

Inasmuch as the structure of the heart is basically the same in all types of a single ventricle, the cardiac findings are similar

The heart at birth is normal in size The rate of enlargement is ordinarily so slow that only when the malformation is compatible with life for several years is there any demonstrable cardiac enlargement Furthermore, a balance is usually established, hence progressive cardiac enlargement, if it occurs at all, occurs extremely slowly

Murmurs are variable During the first months of life a murmur may or may not be present In childhood it is common to hear a precordial *systolic murmur* which may be maximal at the apex Since there is no shunting of blood from an area of high pressure to one of low pressure, the murmur lacks the rasping quality so characteristic of a small ventricular septal defect There may be both a systolic murmur and a low pitched, blurred mid diastolic murmur, thus the murmurs may resemble those of acquired rheumatic heart disease [3] Furthermore, a *gallop rhythm* is frequently heard The transmission of the systolic murmur is, however, not that of a mitral insufficiency and exercise fails to bring out a true crescendo presystolic murmur

The second sound over the pulmonary area is usually accentuated If the pulmonary artery is normally placed, the pulmonary hypertension causes accentuation of the second sound In cases of complete transposition of the great vessels, the aorta occupies the position of the normal pulmonary artery and hence the closure of the aortic valve causes the second sound to be louder to the left of the sternum than to the right Therefore, regardless of the presence or absence of cyanosis, the second sound at the base of the heart to the left of the sternum is usually accentuated It is also frequently reduplicated

Cardiac failure is prone to occur when there is excessive circulation to the lungs As the enlargement occurs extremely slowly, there may be chronic cardiac failure for many years In patients who show no cyanosis, the persistent cardiac *failure may lead to* the diagnosis of an insidious rheumatic infection *Congestion* of the lungs and engorgement of the liver commonly *occur*, and there may be edema and even ascites

The knowledge that the cardiac difficulty dates from infancy or early child hood, *together* with the absence of either history *or* symptoms of acute rheu [illegible] of the nature of the cardiac *difficulty Upon* [illegible]

parently precarious compensation, combined with the [illegible]
sounds, and the absence of any progression of symptoms

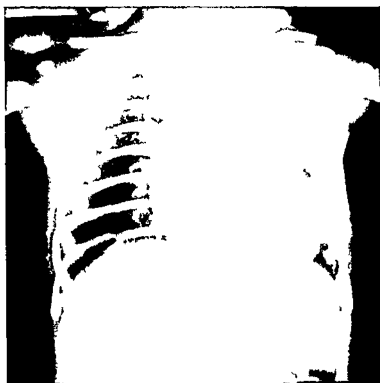
X RAY AND FLUOROSCOPIC FINDINGS

It is the shape of the heart which frequently gives the clue to the diagnosis The fact that the architecture of the heart is the same regardless of the position of the great vessels means that the contour of the heart is the same

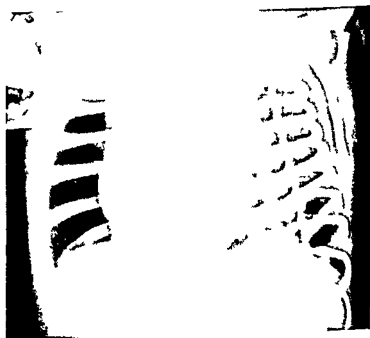
In this malformation there is a striking tendency for overexpansion of the lungs and depression of the diaphragm Consequently the chest is long and nar row and the contour of the heart is also long and narrow

Inasmuch as the rudimentary outlet chamber occupies the position of the [illegible]
bulge in this regio[illegible]
heart is not enlarged, there is prominence of the second curve to the left of the sternum, which suggests that the right ventricle is huge In reality there is no right ventricle, it is only a diminutive chamber It follows that the right ventricle cannot be enlarged The findings in the left anterior-oblique position confirm *this fact* the right ventricle does not project toward the anterior chest wall be yond the margin of the aorta These findings, as shown in Figures xv-4 and 5, demonstrate that the exaggeration of the shadow seen *in* the *anterior posterior position in the region of the* pulmonary conus is not caused by a huge right ven tricle it is in reality due to a rudimentary chamber

Although the contours of the heart are similar when both great vessels arise from the diminutive *outlet* chamber and when only one great vessel arises from the diminutive chamber and the other arises from the common ventricle (com pare Figures xv-4 6 and 7), the hilar shadows may differ greatly When the pulmonary artery arises from the common ventricle there is an excessive blood flow to the lungs, which in turn causes marked pulmonary *congestion* (*see Fig*

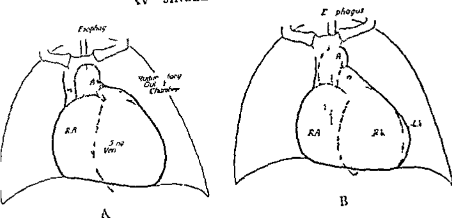


Anterior posterior position

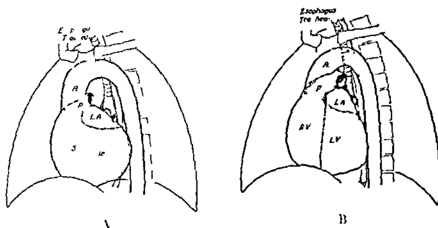


Left anterior-oblique position

FIGURE XX–4 Single ventricle with a rudimentary outlet chamber from which the pulmonary artery arises Infant



ANTERIOR POSTERIOR POSITION



LEFT ANTERIOR-OBLIQUE POSITION

FIGURE XI–7 (A) Single ventricle with a rudimentary outlet chamber and (B) normal heart Infant

ures XI–7, 8, and 11) *When the pulmonary artery is diminutive*, the hilar shadows are minimal Frequently, however, cardiac failure occurs early and by the time the patient comes under observation there may be congestion in the lungs

The contour of the heart changes appreciably with the growth of the individual As the child grows, the diaphragm descends still further and the apex of the heart rotates downward and inward When this occurs, the prominence of the pulmonary conus, which was caused by the rudimentary outlet chamber, disappears the contour of the heart comes to resemble that of a tetralogy of Fallot (compare Figures XI–8, 9 and 10 with Figures VI–17 and 18) or that of a transposition of the great vessels with pulmonary stenosis (see Figure X–26)



FIGURE XV–6 Single ventricle with a rudimentary outlet chamber from which both great vessels arise Infant

If the pulmonary artery arises from the common ventricle, the pulmonary artery is generally large and the pulmonary pressure is increased, consequently hilar pulsations are readily discernible It is the occurrence of greatly increased vascular shadows and a conspicuous hilar dance, combined with a concave curve at the base of the heart to the left of the sternum, which indicates that the pulmonary artery is transposed (see Figures XV–8 and 11) These two x rays show the basic contour of the heart in a child with a single ventricle and posteriorly placed pulmonary artery Nevertheless, the child whose x ray is shown in Figure XV–8 was intensely cyanotic, whereas the one whose x ray is shown in Figure XV–11 was not cyanotic

When the aorta arises from the common ventricle and the pulmonary artery is diminutive in size, there is no prominence of the pulmonary conus, and there is no increase in the pulmonary vascularity Indeed, the lungs may be exceptionally clear Under such circumstances the radiological findings, as shown in Figure XV–12, may closely resemble those of a transposition with pulmonary stenosis Compare Figures XV–12 and X–26

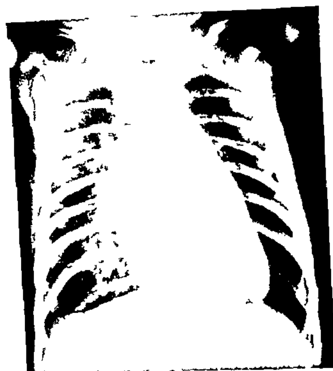


FIGURE XV–7 Single ventricle with a rudimentary outlet chamber from which the aorta arises (same patient as in Figure XV–1) Infant

Examination in the left anterior-oblique position may show that the straight anterior margin of the cardiac silhouette has disappeared by the time the patient reaches late childhood or early adolescence The vascular markings vary with the volume of the pulmonary blood flow, there is, however, always absence of the fullness of the pulmonary conus (see Figure XV–13 and also Figure XV–9) Examination in the right anterior-oblique position contributes little

Occasionally a transposition of the great vessels occurs in combination with pulmonary stenosis in which the aorta, as it arises anteriorly from the rudimentary outlet chamber, arches boldly to the left and causes the pulmonary conus to be extremely prominent and yet the lungs are remarkably clear (see Figure XV–14) This, too, causes the x ray contour of the heart to resemble that of a complete transposition of the great vessels combined with pulmonary stenosis (compare Figures XV–14 and X–27)

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram is subject to wide variation The standard leads



FIGURE XV-8 Single ventricle with a rudimentary outlet chamber from which the aorta arises (same patient as in Figure XV-2) Child

The pulmonary artery arises posteriorly from the main chamber

may show either a right axis deviation or a left axis deviation Although the course of the bundle of His must be grossly misplaced, it is a striking fact that there is usually no demonstrable disturbance of the intraventricular conduction time

Neill and Brink[4] have shown that, even in the presence of a left axis deviation, it is the exception, rather than the rule, for the unipolar precordial leads to show evidence of left ventricular hypertrophy in V_1, and furthermore a patient with a single ventricle may even have a so-called 'septal Q' wave in V_5 or V_6 Indeed, frequently when the standards show a slight left axis deviation, the unipolar precordial leads show evidence of right ventricular hypertrophy in V_6 which causes a deep S wave in all the precordial leads from V_1 to V_6 (see Figure xv-15) In other instances a right axis deviation is combined with left ventricular dominance in the unipolar precordial leads (see Figure xv-16) Thus, the electrocardiogram frequently shows a gross discrepancy between the findings in the standard leads and those in the precordial leads It is, however, possible to have a right axis deviation and right ventricular hypertrophy (see Figure xv-17)



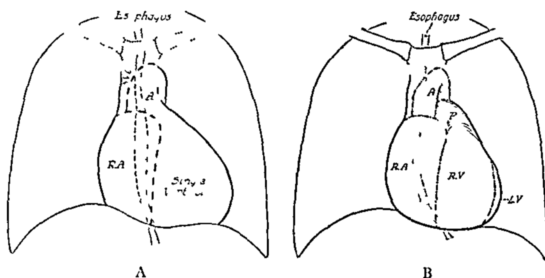
Left anterior-oblique position



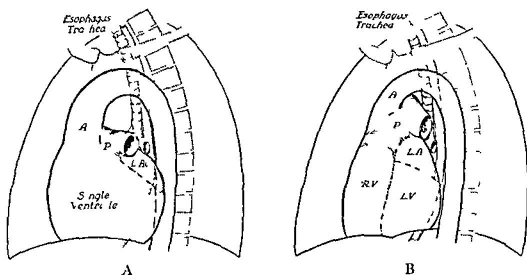
Right anterior-oblique position

FIGURE XV–9 Single ventricle with a rudimentary outlet chamber from which the aorta arises (same patient as in Figure XV–2) Child

The pulmonary artery arises posteriorly from the main chamber



ANTERIOR POSTERIOR POSITION



LEFT ANTERIOR-OBLIQUE POSITION

FIGURE XI-10 (A) Single ventricle with a rudimentary outlet chamber and (B) normal heart Child



Courtesy ... Gl... Gl...

FIGURE XV 11 Single ventricle with a rudimentary outlet chamber from which the aorta arises Child

The pulmonary artery arises posteriorly from the main chamber

SPECIAL TESTS

The oxygen saturation of the arterial blood is never completely normal There is always admixture of the venous and arterial blood in the common ventricle When there is adequate circulation to the lungs and free admixture of the venous and arterial blood in the common ventricle, the oxygen saturation of the arterial blood may be 94 per cent or higher When the pulmonary blood flow is reduced, there is a proportionate reduction in the oxygen saturation of the arterial blood

Polycythemia is proportional to the oxygen unsaturation of the arterial blood Inasmuch as there is always some degree of oxygen unsaturation of the arterial blood the red blood cell count, the amount of available hemoglobin, and the hematocrit reading are usually elevated The height of the red blood cell count varies with the oxygen unsaturation of the arterial blood

In the patient with free admixture of venous and arterial blood in the com



Figure XV-12 Single ventricle with pulmonary stenosis (same patient as in Figure XV-13) Adult

mon ventricle and adequate circulation to the lungs, although polycythemia may not be significant, the red blood cell count is usually a high normal Indeed, the absence of anemia presents a striking contrast to the frail build of the child

If the patient is cyanotic, polycythemia develops early *Once the red blood* cell count, the level of the available hemoglobin, and the hematocrit reading have risen to compensate for the reduction in the oxygen saturation of the arterial blood, polycythemia remains constant over a period of years The patient *does not develop further* hemoconcentration until the pulmonary hypertension becomes extreme

Cardiac catheterization may be of aid in diagnosis The most significant finding on cardiac catheterization is the marked increase in the oxygen content of the blood in the common ventricle as compared with that in the right auricle Bing[*] considered that an increase of 5 volumes per cent in the oxygen content of the blood of the right ventricle, compared with that of the right auricle, is indicative of a common ventricle Such a finding is strong confirmatory evidence of a common ventricle, but such an increase can occur only if the difference between



Left anterior-oblique position



Right anterior-oblique position

FIGURE XV–13 Single ventricle with pulmonary stenosis (same patient as in Figure XV–12) Adult



Teleoroentgenogram



Angiocardiogram

FIGURE XV-14 Single ventricle with transposition of the great vessels, the aorta arising far to the left Infant

the oxygen content of the arterial blood and that of the venous blood is greater than 5 volumes per cent When there is tremendous pulmonary blood flow, the oxygen content of the venous blood may be so high that there is less than a 5 volume percentage difference between the arterial and venous blood Conversely, when there is severe pulmonary stenosis, the pulmonary blood flow may be so meager that the arterial oxygen saturation is too low to have a 5 volume percentage difference Furthermore, if a single ventricle occurs in combination with a single auricle, the increase in the oxygen content of the blood will occur in the auricle, and the oxygen content of the blood in the auricle and in the ventricle will be the same Therefore the finding of a marked increase in the oxygen content of the blood in the ventricle is suggestive evidence of a single ventricle, but its absence does not exclude the diagnosis

Angiocardiography shows simultaneous opacification of the aorta and the

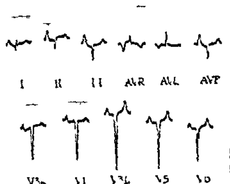


FIGURE XV-15 Single ventricle with pulmonary stenosis Adult
Note the deep S in all precordial leads left ventricular dominance in V_1 and right ventricular dominance in V_6

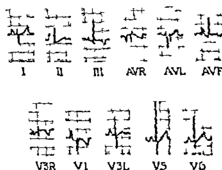


FIGURE XV-16 Single ventricle
Note the right axis deviation and the normal left ventricular dominance

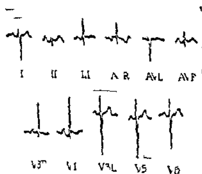


FIGURE XV-17 Single ventricle
Note the right axis deviation and right ventricular hypertrophy

pulmonary artery, it also clearly delineates the course of the aorta (see Figure xv-14) Furthermore, the dye promptly fills the entire ventricle, which usually has a smooth outline in contrast to the normal trabeculation of the right ventricle Such a contour does not exclude the possibility of a huge right ventricle, nor yet the absence of the right ventricle and the existence of a single ventricle, the left Nevertheless, when angiocardiography is undertaken to determine whether or not there is a single ventricle or two ventricles, the visualization of the right ventricle and later of the left ventricle demonstrates the existence of two ventricles

DIAGNOSIS

The diagnosis is based upon a combination of clinical, cardiological, and radiological findings, these vary with the age of the patient

In early infancy there is usually cyanosis of varying intensity, and a small heart with slight fullness of the pulmonary conus in the anterior posterior position, but no enlargement of the right ventricle in the left anterior oblique position Cyanosis may be so intense and the arterial oxygen saturation may be so low that the infant is in danger of dying from anoxemia, or cyanosis may be so slight that no cardiac abnormality is suspected and the condition is misdiagnosed as pneumonia

When there is pulmonary atresia, cyanosis is intense and the infant suffers from paroxysmal dyspnea, the heart is slightly enlarged and has a square contour similar to that of tricuspid atresia and the lungs are clear The discrepancy in the electrocardiogram between the standard leads and the unipolar precordial leads indicates that the condition is neither a tetralogy of Fallot nor a tricuspid atresia

In childhood there are at least three distinct clinical syndromes (1) when the pulmonary blood flow may be adequate or excessive with free admixture of venous and arterial blood in the common ventricle, (2) when the great vessels may be transposed and the pulmonary circulation is adequate but the occurrence of an abnormality of the tricuspid valve directs the venous blood into the aorta, (3) when the pulmonary artery may be small or the pulmonary valve stenotic and consequently pulmonary blood flow is reduced

In the first instance, *when the pulmonary blood flow is adequate or excessive and cyanosis is absent,* the diagnosis is based upon the finding of a patient of frail build with slight cardiac enlargement, with a precordial systolic murmur, and with a blurred mid diastolic murmur at the apex Fluoroscopy reveals in

creased hilar shadows The red blood cell count is normal but the oxygen saturation of the arterial blood is slightly reduced

In the second instance, *when cyanosis is intense and the pulmonary blood flow is adequate,* there is always polycythemia The heart is slightly enlarged and the hilar shadows are increased and may show a conspicuous *hilar dance* but the shadow at the base of the heart is concave The patient is less limited than might be expected from the intensity of the cyanosis When the pulmonary artery is huge, this malformation closely simulates that of a complete transposition of the great vessels with an enormous pulmonary artery (see Chapter X, Section B)

In the third instance, *when cyanosis is intense and the pulmonary blood flow is reduced* polycythemia develops early, the heart is only slightly enlarged but has a concave curve at its base to the left of the sternum, murmurs are variable, the pulmonic second sound is accentuated but the lungs are extremely clear, and the vascular markings are reduced

The infant does better than is anticipated from the intensity of the cyanosis The child can often play all day without undue fatigue In such instances the x ray may be similar to that of a tetralogy of Fallot but the child does not squat when tired The electrocardiogram usually offers a clue to the diagnosis, as there is frequently a striking discrepancy between the axis deviation in the standard leads and the evidence of left or right ventricular dominance in the unipolar precordial leads

DIFFERENTIAL DIAGNOSIS

In the absence of cyanosis a single ventricle requires differentiation from pneumonia and rheumatic heart disease In the presence of cyanosis, the condition requires differentiation from a tetralogy of Fallot from a tricuspid atresia, from a complete transposition of the great vessels with two ventricles and a posteriorly placed dilated pulmonary artery, and occasionally from a truncus arteriosus with a moderate reduction in the pulmonary blood flow

Pneumonia is a common error in diagnosis in infants When there is no murmur and the heart is of normal size, the occurrence of slight cyanosis and dyspnea combined with extensive pulmonary congestion, is strongly suggestive of pneumonia The failure to demonstrate pathogenic organisms in cultures from the nose and throat, combined with the contour of the heart in the x ray, offers a clue to the correct diagnosis Indeed, if the pathologist implies that the clinician has erroneously diagnosed pneumonia in a young infant who was

found to have a congenital malformation of the heart, the possibility of a bilocu late or triloculate heart should be immediately entertained

Rheumatic heart disease may be erroneously diagnosed in children who show no cyanosis Although the murmurs may superficially resemble those of a rheu matic myocarditis, careful analysis of murmurs will show that they are not char acteristic of that disease The systolic murmur is not well transmitted to the axilla The early diastolic murmur at the apex does not resemble a presystolic murmur The absence of a rheumatic history in a child with symptoms of long duration is also significant The red blood cell count, the amount of available hemoglobin, and the hematocrit reading are usually normal or slightly elevated, whereas with a long standing rheumatic infection anemia is the rule Simple awareness of the possibility aids in the diagnosis of a single ventricle Fluoroscopic examination corroborates the diagnosis Determination of the oxygen saturation of the arterial blood will reveal that it is not fully saturated Cardiac catheterization will show definite evidence of admixture of venous and arterial blood in the common ventricle

When the patient shows persistent cyanosis, x ray or fluoroscopy will show whether the pulmonary blood flow is increased or decreased When the pulmonary blood flow is increased, a single ventricle calls for differentiation from complete transposition of the great vessels with a dilated pulmonary artery, when decreased, a single ventricle requires differentiation from a tetralogy of Fallot, tricuspid atresia, a truncus arteriosus, and a complete transposition of the great vessels with pulmonary stenosis

Transposition of the great vessels combined with a greatly dilated pulmo nary artery and an intact ventricular septum gives identically the same x ray or fluoroscopic findings as does the Lambert heart The patient is usually more severely incapacitated if there are two ventricles because of the greater difficulty in the admixture of oxygenated and venous blood Cardiac catheterization is generally necessary in order to differentiate the two conditions Although the aorta may be readily entered, the catheter usually also slips into the common ventricle, in which the oxygen content of the blood will be far higher than it is in the right ventricle or in the aorta

Complete transposition of the great vessels with an auricular septal defect may occasionally show a tremendous increase in the oxygen content of the blood in the right ventricle and may be extremely difficult to differentiate from a Lambert heart Angiocardiography may be of aid by demonstration of an auricular defect, and also will usually demonstrate the existence of two ventricles and the late opacification of the pulmonary arteries

A tetralogy of Fallot may be easily confused with a single ventricle combined with *pulmonary stenosis*, primarily because of the contour of the heart A history of freedom from attacks of paroxysmal dyspnea and of absence of squatting is against the diagnosis of a tetralogy of Fallot Furthermore, on physical examination the systolic murmur usually is not as loud or as harsh as in a tetralogy of Fallot and the pulmonic second sound is accentuated The electrocardiogram also generally aids in the differentiation of the two conditions

A tricuspid atresia can usually be differentiated by the electrocardiogram, occasionally cardiac catheterization is necessary to determine whether or not a ventricle can be entered from the right auricle

A truncus arteriosus with moderately reduced pulmonary blood flow may be considered because of the absence of squatting and the relatively good exercise tolerance The continuous murmur audible over the lungs readily differentiates a truncus arteriosus from a single ventricle A truncus arteriosus may, however, be associated with a single ventricle, but if so, the single ventricle is mainly of academic interest, as all the blood from the common ventricle is pumped into the truncus arteriosus (see Chapter xiv)

Transposition of the great vessels combined with pulmonary stenosis is probably the most difficult of all conditions to differentiate from a single ventricle when the stenosed pulmonary artery arises from the common ventricle Regardless of whether the aorta arises normally from the mid portion of the right ventricle or whether it arches boldly to the left, the condition may simulate that of a single ventricle. Indeed, the demonstration of two ventricles by angiocardiography is virtually the only way to differentiate the two conditions with certainty

COMMONLY ASSOCIATED ANOMALIES

A single ventricle may occur in combination with tricuspid atresia Furthermore, this combination of anomalies may occur with or without a transposition of the great vessels, and with or without pulmonary stenosis Functionally the condition is closely similar to a tricuspid atresia and an underdeveloped right ventricle There are two auricles and a single ventricle The blood from the right auricle can leave only by way of the left auricle and thence the admixture of venous and arterial blood enters the common ventricle and is pumped out to both the systemic and pulmonary circulations

In the presence of a single ventricle, the blood usually circulates freely in the common chamber and less difficulty is encountered in the expulsion of blood from the rudimentary outlet chamber than through a small defect into a rudi

mentary right ventricle, hence the condition is more frequently compatible with life for a number of years than when tricuspid atresia is combined with a rudimentary right ventricle Nevertheless, the two conditions merge into one another It may be difficult even at autopsy to decide which condition is present

The electrocardiogram provides the principal clinical clue to the differentiation of the two anomalies Usually in a single ventricle the unipolar precordial leads do not show the evidence of left ventricular hypertrophy in V_1 which is the rule in tricuspid atresia Neill and Brink[4] have, however, reported two cases in which the electrocardiograms were closely similar to that of tricuspid atresia

A single ventricle may also occur in combination with mitral atresia as illustrated *in Figure* XI–*18* When the *mitral* valve is atretic, a gross defect *in the auricular septum is inevitable, as such an opening is the only way by which the* blood can escape from the left auricle The smaller the defect in the auricular septum, the greater is the enlargement of the left auricle The occurrence of a single ventricle in combination with mitral atresia causes the pulmonary pressure to be excessively high because the pulmonary resistance is increased both by the high pressure in the left auricle (see Chapter XXX) and by the high pressure under which the blood is ejected to the lungs from the common ventricle Consequently pulmonary hypertension is extreme and pulmonary congestion is severe

TREATMENT

When a single ventricle occurs in combination with pulmonary stenosis, a systemic pulmonary anastomosis increases the circulation to the lungs and lessens cyanosis but may lead to progressive cardiac enlargement A number of patients with the single ventricle and pulmonary stenosis die of cardiac failure within six to eight months after operation, others develop cardiac enlargement more slowly and may live for eight to ten years or longer Nevertheless, some patients with a single ventricle are as greatly benefited by this operation as are those with a tetralogy of Fallot

If the primary difficulty is due to excessive pulmonary blood flow, the production of moderate pulmonary stenosis may be of benefit to the patient Obviously it is desirable that the pulmonary stenosis be sufficient to break the pulmonary pressure but not so great as to decrease pulmonary blood flow to the extent that it will produce cyanosis Muller and Dammann[6] have developed a method for decreasing the size of the pulmonary artery by means of a "tucking procedure Such an operation may be of great value to the patient with a single ventricle

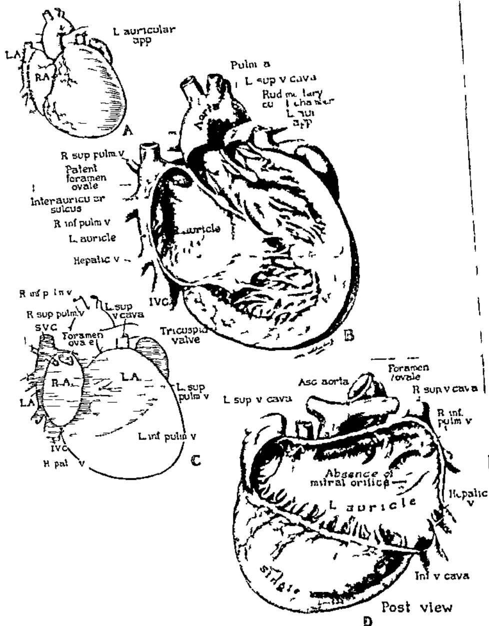


FIGURE XV 18 Single ventricle and mitral atresia

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from the rudimentary outlet chamber and the insertion of the chordae tendineae of the tricuspid valve are such that the blood from the right auricle is directed into the rudimentary outlet chamber, the resultant malformation is known as a Lambert heart

Inas [illegible] all the blood from both auricles flows in th [illegible] is [illegible] of the blood [illegible] which reaches the lungs for oxygenation If the total volume [illegible] to the lungs is small, the volume of oxygenated blood returned from the lungs is proportionally small and the oxygen content of the blood in the common ventricle is low Unless there is pulmonary stenosis, there is always pulmonary hypertension

The presence or absence of cyanosis depends upon the oxygen content of the blood in the common ventricle and the amount of venous blood directed into the aorta Usually there is free admixture of venous and arterial blood in the common ventricle and cyanosis is minimal or absent In the Lambert heart, although the oxygen content of the blood in the common ventricle is relatively high, the oxygen content of the blood in the rudimentary outlet chamber is low little oxygenated blood is directed into the aorta cyanosis is intense

A high red blood cell count is the rule In the absence of cyanosis the only evidence of polycythemia may be the persistence of the red blood cell count at a high normal level in a patient who looks chronically ill and suffers from repeated pulmonary infections When the patient shows persistent cyanosis, polycythemia is the rule

Clubbing is directly proportional to the polycythemia

The exercise tolerance of the individual varies with the oxygen saturation of the arterial blood nevertheless, in cyanotic patients it is better than is anticipated from the intensity of the cyanosis

The heart is usually but slightly enlarged The second sound is generally accentuated Murmurs are variable and may simulate those of a poorly functioning rheumatic heart

The x ray and fluoroscopic findings frequently give a clue to the diagnosis In infancy there is fullness of the pulmonary conus but no evidence of right ventricular enlargement in the left anterior-oblique position In older children, when the pulmonary artery is of small caliber and the lung fields are clear, the

If a child has a small pulmonary artery and suffers from reduced pulmonary blood flow under relatively high pulmonary pressure, it may be difficult, if not impossible, to help him by operation An anastomosis will only function provided the pulmonary pressure is lower than the systemic pressure Corrective surgery would necessitate the construction of a septum within the ventricles in order to reduce the pressure with which blood is directed to the lungs In addition, if there is but a single atrioventricular valve, it will be necessary to reconstruct the atrioventricular orifice so that there is both a mitral and a tricuspid valve

If the patient has a Lambert heart, the creation of an auricular septal defect may increase the amount of oxygenated blood directed to the aorta but it will not alleviate the pulmonary hypertension

PROGNOSIS

The prognosis is guarded *In the absence of cyanosis and excessive pulmonary blood flow*, the patient is very susceptible to pneumonia, which is a constant menace to life *In the presence of cyanosis and excessive pulmonary blood flow*, the increasing pulmonary hypertension increases the volume of blood directed to the systemic circulation and consequently the condition of the patient remains relatively stationary for ten or twenty years Nevertheless, in both instances the pulmonary hypertension will eventually render the condition incompatible with life

In the presence of pulmonary stenosis there is always cyanosis Any operation which increases the circulation to the lungs may help Although some patients are greatly helped by operation, it must be admitted that the human circulation and metabolism are based on a four chambered heart and the separation of the two circulations A single ventricle is not an efficient mechanism and consequently the prognosis is guarded

SUMMARY

A single ventricle with a rudimentary outlet chamber represents an extremely early arrest in the development of the heart Instead of two ventricles, there is but a single ventricle into which both the mitral and tricuspid valves open In the position normally occupied by the outflow tract of the right ventricle, there is a rudimentary chamber from which one or both the great vessels arise The great vessels may or may not be transposed When the aorta arises

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contour of the heart may closely simulate that of a tetralogy of Fallot When the pulmonary artery is of normal size and posteriorly placed, the hilar vascularity is increased and there is a conspicuous hilar dance

The electrocardiogram is subject to great variation There may be a right or a left axis deviation, but usually there is a discrepancy between the axis deviation in the standard leads and the evidence of hypertrophy in the unipolar precordial leads

Cardiac catheterization characteristically shows a marked increase in the oxygen content of the blood in the common ventricle in comparison with that in the right auricle The systolic pressure in the common ventricle is always the same as the systolic pressure in the systemic circulation, consequently, unless there is pulmonary stenosis, there is always pulmonary hypertension

Angiocardiography shows simultaneous opacification of the aorta and the pulmonary artery but is not of great aid in the diagnosis of the structure of the ventricle

The diagnosis is based upon the clinical findings and the contour of the heart in the x ray combined with electrocardiographic findings of a discrepancy between the axis deviation in the standard leads and the ventricular dominance in the precordial leads The diagnosis may be substantiated by cardiac catheterization

In the absence of cyanosis, the condition must be differentiated from pneumonia in infancy and from rheumatic heart disease in childhood If there is persistent cyanosis, the malformation may resemble a tetralogy of Fallot The Lambert heart requires differentiation from a complete transposition of the great vessels with a dilated pulmonary artery and occasionally from a truncus arteriosus

Treatment is not satisfactory If cyanosis is absent and the pulmonary blood flow excessive, a tuck in the pulmonary artery may be of great help If cyanosis is intense and the pulmonary blood flow excessive, the creation of an auricular defect may help When the pressure is the same in both great vessels, an anastomosis will not function, hence it may be impossible to help a child with persistent cyanosis and a relatively small pulmonary artery

The prognosis is guarded Many infants with intense cyanosis and minimal pulmonary blood flow die within the first few months Patients with a moderate reduction in pulmonary blood flow may live to childhood Most patients with adequate pulmonary blood flow, with or without cyanosis, survive to adult life

rare The sequence of events is illustrated by Case ...

Cerebral arteriovenous aneurysms unfortunately, are not rare Usually they are small and asymptomatic until early adult life The outstanding physical finding is a localized bruit Such aneurysms may bleed before they finally rupture or they may remain entirely unsuspected until the young adult suddenly develops a severe headache and dies of a massive cerebral hemorrhage

The frequency of congenital cerebral arteriovenous aneurysms in patients with malformations of the heart is difficult to ascertain Systolic bruits over the skull are extremely common in patients with cardiac malformations Certainly some of these bruits are the result of the bone transmission of cardiac murmurs Continuous murmurs over the skull are not infrequently heard in patients with persistent cyanosis Many such patients are entirely asymptomatic Inasmuch as extensive investigation of cerebral abnormalities is not without risk, it has not been our policy to undertake such studies in an asymptomatic person unless there were some localizing signs Nevertheless, in a number of instances a continuous murmur over the skull may be indicative of a cerebral arteriovenous aneurysm[4]

Coronary arteriovenous aneurysms are fundamentally similar to other congenital arteriovenous aneurysms but as they represent an anomaly of the coronary arteries they are discussed in Chapter XXIX

B *Pulmonary Arteriovenous Aneurysms*

Arteriovenous aneurysms in the lungs have been reported under various names hemangiomas of the lung, cavernous angiomas, and pulmonary arteriovenous aneurysms The first unequivocal case is credited to Rode,[5] in 1938 The condition was reviewed and reported as a new surgical disease by Watson[6] in 1947 Two years later the radiological findings were reported by Grishman et al[7] For a detailed review the reader is referred to the reports by Sloan and Cooley[8,9] Almost all arteriovenous aneurysms are congenital in nature Some aneurysms undoubtedly increase in size as the patient grows and therefore become clinically manifest in late childhood or early adult life In former years the condition was frequently misdiagnosed as a congenital malformation of the heart and considered inoperable With the advent of cardiac surgery, many of these patients, even though asymptomatic, have sought medical attention and have been helped

NATURE OF THE MALFORMATION

The malformation consists of an abnormality in the pulmonary vascular bed

CHAPTER XVI

ARTERIOVENOUS ANEURYSMS

AN arteriovenous aneurysm is defined as a blood containing tumor or cavity with both a systemic and a venous connection Although some are the result of trauma, this chapter is concerned with those which are congenital in origin It seems probable that many, if not all hemorrhagic telangiectases, hemangiomas, and cavernous angiomas are fundamentally arteriovenous aneurysms Indeed, the frequent association of hemorrhagic telangiectases and pulmonary arteriovenous aneurysms suggests that both are essentially of the same nature [1]

Arteriovenous aneurysms may occur anywhere in the systemic or the pulmonary circulation They may occur in the extremities, in the brain, or even in the myocardium, and they also occur in the lungs When it is recalled that arteries and veins develop independently, it is not surprising that anomalous connections between the arteries and veins may occur anywhere in the body, and, furthermore, if the development is such that an arteriovenous aneurysm occurs in one place, others may occur in other locations Therefore, although there may be only a single arteriovenous aneurysm, frequently they are multiple The size of the anomalous connection varies greatly, it may be large or small

A *Systemic Arteriovenous Aneurysms*

Systemic arteriovenous aneurysms vary in size Some are minute and entirely asymptomatic Others may be large When an arteriovenous aneurysm occurs in the systemic circulation, blood is shunted under systemic pressure into a vein, consequently the volume of the shunt is usually large The blood so shunted is fully oxygenated *It follows that a large volume of fully oxygenated blood is re*turned to the right auricle, thence it flows to the right ventricle and is re circulated through the lungs The work of the right side of the heart is increased A large arteriovenous fistula may greatly increase the work of the heart and may lead to cardiac failure due to the high output of the heart

The hereditary nature of hemorrhagic telangiectases was described by Osler in 1901 and by Weber[3] in 1907 This syndrome is now known as Rendu Osler Weber's disease Such telangiectases may occur in almost any organ of the body The problem is primarily medical and hence beyond the scope of this book

Systemic arteriovenous aneurysms, congenital in origin, which are so large

culation The shunt, however, occurs in the lungs Consequently it occurs in an area of low pressure There is no tendency for the patient to develop pulmonary hypertension The only effect on the circulation is that the arterial blood is not fully oxygenated and the work of the heart is increased

CLINICAL FINDINGS

The patient frequently remains asymptomatic for many years If he has a complaint it is usually that his color is not normal

Cyanosis ordinarily dates from birth The intensity of the cyanosis depends upon the size and number of the arteriovenous aneurysms in the lungs Occasionally, although some degree of oxygen unsaturation of the arterial blood has been present from birth the volume of reduced hemoglobin in the circulating blood is insufficient to cause visible cyanosis This is notably true in infancy, because anemia is common

Polycythemia develops secondary to the oxygen unsaturation of the arterial blood With the development of polycythemia, cyanosis becomes more readily apparent

Clubbing of the extremities may eventually appear

Dyspnea is not striking but there may be exertional dyspnea Although some blood is shunted through the lungs without being oxygenated, a large portion of the pulmonary circulation is normal hence the patient is able to increase his pulmonary blood flow with exercise Consequently dyspnea is minimal The patient s exercise tolerance is better than might be expected from the degree of cyanosis

Anemia may be the presenting complaint Many patients with pulmonary arteriovenous aneurysms also have systemic arteriovenous aneurysms or hemorrhagic telangiectases If these are located in the gastrointestinal tract or in other mucous membranes, persistent oozing and bleeding may lead to severe anemia

Hemorrhagic telangiectases and hemangiomas are extremely common in patients with pulmonary arteriovenous aneurysms Therefore a search for these should always be made The converse is also true The presence of a systemic arteriovenous aneurysm should suggest the possibility that the cyanosis may be due to an aneurysm in the pulmonary circulation

Epistaxes are common They are in all probability due to minute arteriovenous aneurysms in the nose

Hemoptyses are a serious and late complication and may vary from repeated small hemorrhages to a massive fatal hemorrhage

of such a nature that there is free and broad communication between some of the arteries and veins within the lungs There may be single or multiple aneurysms, they may be large or small, such aneurysms may be localized in one lobe or scattered throughout both lungs The aneurysm usually involves the smaller branches of the pulmonary artery, which, instead of subdividing and branching into the capillary bed, open into a sacular cavity and this in turn communicates directly with the veins Frequently there are numerous cavities of various sizes, occasionally the lung is riddled with small cavities and often both lungs are involved In rare instances an aneurysm may occur between the bronchial artery and the pulmonary vein or may even occur in an anomalous artery leading to the lungs, as in the case reported by W L Watson,[9] in which there was an arteriovenous aneurysm between an aberrant artery from the thoracic aorta and what appeared to be a pulmonary vein The author has seen one patient in whom the anomalous branch from the pulmonary artery opened directly into the left auricle (see Case xvi–2 at end of chapter)

Pulmonary arteriovenous aneurysms tend to increase in size just as other aneurysms do The increase in size, however, occurs more gradually in a low pressure area than in a high pressure area For this reason it may require years for the increase in the size of aneurysms to become apparent

COURSE OF THE CIRCULATION

The blood from the right auricle flows into the right ventricle and is pumped out in the normal fashion to the lungs Part of the blood circulates normally through the normal lung, where it is oxygenated, and returned in the normal fashion to the left auricle A varying volume of blood passes directly from the pulmonary artery through the arteriovenous aneurysm to the pulmonary veins Consequently a mixture of oxygenated and unoxygenated blood is returned by the pulmonary veins to the left auricle This admixture of venous and arterial blood flows into the left ventricle, is pumped out through the aorta to the systemic circulation, and is returned to the right auricle in the normal manner There the cycle starts again (see Diagram xvi–1)

PHYSIOLOGY OF THE MALFORMATION

The shunting of the venous blood from the pulmonary artery into a pulmonary vein without passing through the capillaries prevents the oxygenation of the blood in that area of the lung Venous blood is returned to the left auricle and thence it flows to the left ventricle and is pumped out into the systemic cir

culation The shunt, however, occurs in the lungs Consequently it occurs in an area of low pressure There is no tendency for the patient to develop pulmonary hypertension The only effect on the circulation is that the arterial blood is not fully oxygenated and the work of the heart is increased

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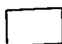
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Hemoptyses are a serious and late complication and may vary from repeated small hemorrhages to a massive fatal hemorrhage

DIAGRAM XVI-1

Head and upper extremities
S.V.C.
P.A.
P.V.
R
L
Aorta
Pul. A.
Pul. V.
Pulmonary art.
L.A.
R.A.
L.V.
R.V.
Descending Aorta
I.V.C.
Trunk and lower extremities


Arterial blood (fully saturated)


Small admixture of venous blood
No visible cyanosis


Venous and arterial blood
Cyanosis visible


Venous blood

Diagram XVI–1

Multiple arteriovenous aneurysms

This malformation concerns the pulmonary arteries and pulmonary veins, the heart is itself normally formed The major part of the pulmonary vascular bed is normal but scattered throughout the lung there are arteriovenous aneurysms connecting some of the smaller branches of the pulmonary arteries with the pulmonary veins

The blood from the right auricle flows into the right ventricle and thence is pumped out into the pulmonary artery to the lungs Part of the blood circulates through the normal portions of the lungs where it is oxygenated, and is returned in the normal manner by the pulmonary veins to the left auricle. Part of the blood which is pumped to the lungs flows through an arteriovenous aneurysm The blood which circulates through such an aneurysm never reaches the pulmonary capillaries and hence is returned to the left auricle unoxygenated Thus a mixture of oxygenated and venous blood flows through the pulmonary veins to the left auricle thence the blood flows into the left ventricle and is pumped out through the aorta to the body and returned by the superior vena cava and the inferior vena cava in the normal manner to the right auricle There the cycle starts again

Clinical diagnosis Cyanosis is persistent and increases as the arteriovenous aneurysm increases in size. The patient is usually asymptomatic The heart is normal in size No murmurs are audible over the precordium but a continuous murmur is generally audible over the aneurysm If the aneurysms are large or increase in size, the patient is in danger of severe, even fatal pulmonary hemorrhages

Cerebral disturbances may occur

Transitory neurological disturbances are relatively frequent These may be due to anoxemia, polycythemia, minute thromboses, or to bleeding from minute cerebral aneurysms

Cerebral thromboses may occur in conjunction with small cerebral arteriovenous aneurysms

CARDIAC FINDINGS

The heart is generally normal in size Usually no murmurs are audible over the precordium The *pulmonic second sound* may be slightly accentuated A *soft continuous murmur* is usually audible over the area of the lung where there is a relatively large arteriovenous aneurysm

X RAY AND FLUOROSCOPIC FINDINGS

The x ray generally gives the clue to the diagnosis [8, 9] Although the heart is essentially normal in size and shape, an abnormal shadow is usually visible somewhere in the lungs (see Figure xvi–1) These abnormal shadows can frequently be seen upon fluoroscopy, indeed, intrinsic pulsations of the shadows may even be discernible When an arteriovenous aneurysm is suspected but not clearly evident, the patient should be carefully examined in all positions, because the aneurysm may be hidden behind the cardiac shadow

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram usually shows a balanced axis and no evidence of right ventricular hypertrophy

SPECIAL TESTS

The oxygen saturation of the arterial blood is reduced Nevertheless, the degree of unsaturation is less than is anticipated from the intensity of the cyanosis Furthermore, the arterial oxygen saturation falls but slightly, if at all, with exercise

Cardiac catheterization studies are frequently misleading The Fick principle is no longer valid because the blood returned to the left auricle is not fully saturated The pressures are normal, the resistance in the pulmonary vascular bed is normal [10]

Angiocardiography is of great diagnostic aid, it not only opacifies the princi



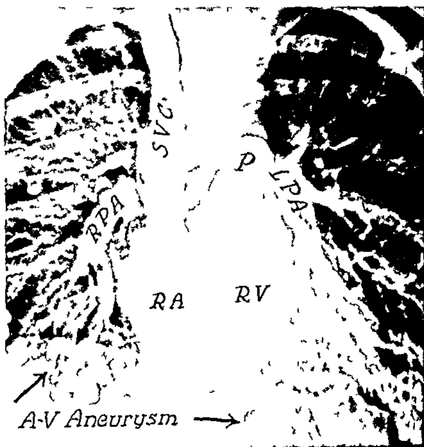
Figure XVI–1 Multiple pulmonary arteriovenous aneurysms (same patient as in Figure XVI–2) Adult

pal *arteriovenous aneurysm* but may also demonstrate additional unsuspected aneurysms (see Figure XVI–2) Angiocardiography is, however, not without danger, as the dye is pooled in the aneurysms Deaths have been reported from this procedure[11]

Laminograms are of great aid Such films give an indication of the size and position of the aneurysm and may reveal the tortuous vessels in and around it Planograms do not cover the entire lung and consequently other aneurysms may be missed Nevertheless, planography is a simple and safe procedure which aids in localization of the mass

DIAGNOSIS

The diagnosis should be suspected in a patient with persistent cyanosis and no limitation of activity and no complaints Physical examination reveals no cardiac abnormality A localized *continuous murmur* over the lungs is strong presumptive evidence in favor of the diagnosis The diagnosis is substantiated by



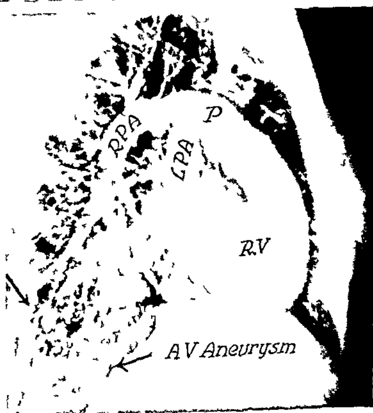


FIGURE XVI-2 Multiple pulmonary arteriovenous aneurysms (same patient as in Figure XVI-1) Adult

ray or fluoroscopy and may be confirmed by an angiocardiogram or by a aminogram

DIFFERENTIAL DIAGNOSIS

The condition requires differentiation from congenital methemoglobinemia, from anomalous communication between the pulmonary artery and the left auricle, and also from congenital malformations of the heart, notably a truncus arteriosus, or localized stenoses in the branches of the pulmonary artery

Congenital methemoglobinemia is to be suspected in a patient with an absolutely normal heart, normal lungs, and persistent cyanosis which dates from birth The usual story is that the abnormal color was noted at birth Nevertheless the infant gains and thrives so well that the cyanosis causes little or no concern Even if a physician is consulted, the infant is doing so well that further investigations are seldom advised It is the normality of growth and development in the presence of persistent cyanosis that gives the clue to the diagnosis Occasionally the condition is familial and the knowledge that other members of the family have also shown an abnormal color aids in the diagnosis

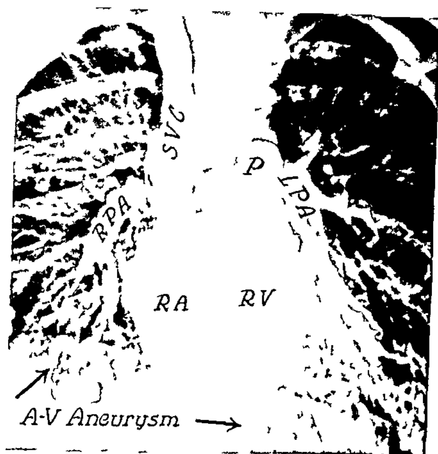
[illegible] ingestion of nitrites [illegible] well water in a [illegible] baby s formula[1, 13]

The differentiation of cyanosis caused by methemoglobin from that caused by the presence of reduced hemoglobin in the circulating blood can easily be determined by the following test A drop of the patient s blood is placed on a piece of paper beside a drop of one s own blood When the two samples are stirred, the normal venous blood becomes bright red whereas the blood which contains methemoglobin is unable to combine with the oxygen in the air and therefore retains its dark color[14]

The diagnosis is definitively established by spectroscopic analysis of the blood and by the demonstration of the band characteristic of methemoglobin Cyanosis can be relieved by the injection of methylene blue and the patient can be cured by the continuous use of massive doses of ascorbic acid

Abnormal vascular communication between the pulmonary artery and the left auricle may simulate the picture of a pulmonary arteriovenous aneurysm (see Case XVI-2 at end of chapter)

A truncus arteriosus also causes persistent cyanosis and a continuous murmur over the lung Examination of the heart usually reveals a harsh systolic murmur and accentuation of the second sound over the base of the heart The contour of



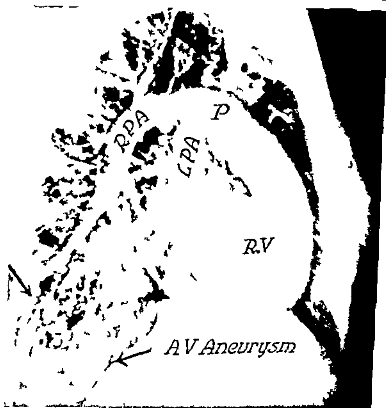


FIGURE XVI-2 Multiple pulmonary arteriovenous aneurysms (same patient as in Figure XVI-1) Adult

Pulmonary arteriovenous aneurysms are *frequently multiple* Inasmuch as venous blood from the pulmonary artery is shunted into the pulmonary vein without passing through the capillaries of the lungs the blood returned to the left auricle is not fully saturated Consequently the arterial blood is not fully saturated

The patient is cyanotic but usually asymptomatic The heart is normal in size The outstanding finding is a localized continuous murmur audible over the lung at the site of the aneurysm The aneurysm is frequently visible upon x ray or fluoroscopy and can almost invariably be delineated by angiocardiography or laminography

The condition calls for differentiation from congenital methemoglobinemia and other conditions which cause a continuous murmur over the lungs, notably peripheral pulmonary stenoses and a truncus arteriosus

The condition can be corrected by surgery

Prognosis is guarded but can be greatly improved by surgery

Unusual Illustrative Cases

CASE XVI-1 B S (Harriet Lane Home No A-98289) White male. Admitted at thirty six hours of age because of a mass in the right temporal region and severe cardiac failure

History The baby was born at the Lutheran Hospital He cried immediately and breathed well and then developed apnea He was given positive pressure oxygen and respiration was restored Shortly after birth the nurse noted a mass in the right temporal region Examination of the mass revealed a thrill and a bruit This mass rapidly increased in size Respiration became rapid Because of the dyspnea an x ray of the heart was taken at eighteen hours of age this revealed cardiac enlargement The baby was given digitalis

Physical examination The infant was dyspneic and cyanotic The respiratory rate was 200 per minute The heart was greatly enlarged and there was a gallop rhythm The liver extended to the umbilicus There were rales in the lungs

Over the right temporal region there was a large, soft pulsating mass approximately 5 cm in diameter In the center of this area there was no bony structure consequently the mass lay beneath the skin Around the periphery of the mass a delicate rim of the bone could be palpated A thrill was palpable over the mass and on auscultation there was a continuous bruit with a systolic accentuation Both the murmur and the thrill could be eliminated by pressure over the carotid artery

Diagnosis The infant obviously had a large arteriovenous aneurysm involving the carotid artery and high-output cardiac failure

the heart and the electrocardiogram are also abnormal Cardiac catheterization shows that the aorta arises in part or entirely from the right ventricle (see Chapter xiv)

Localized constrictions in one or more of the branches of the pulmonary artery are also a possible cause of a continuous murmur over the lungs The condition by itself does not produce cyanosis Nevertheless, peripheral pulmonary stenoses frequently occur in combination with some malformation of the heart which does cause cyanosis Therefore if a continuous murmur is due to peripheral stenoses and there is cyanosis, examination of the heart should reveal some abnormality

TREATMENT

Excision of the affected area offers the only possible cure Lobectomy or pneumonectomy is indicated if the major pathology lies in one lung, as there is real danger of serious, even fatal, hemorrhage

PROGNOSIS

Pulmonary arteriovenous aneurysms may be compatible with life for many years If the aneurysms are large and localized, or if the patient has had hemoptyses, operation is clearly indicated Provided the mortality rate from lobectomy or pneumonectomy is low, operation is indicated for any patient with a pulmonary arteriovenous aneurysm sufficiently large to cause cyanosis, as there is always the potential danger of rupture of the aneurysm

SUMMARY

An arteriovenous aneurysm is a blood containing tumor or cavity with both an arterial and a venous connection It seems probable that hemorrhagic telangiectases, hemangiomas, and cavernous angiomas are all basically arteriovenous aneurysms Such arteriovenous aneurysms may occur in either the systemic or the pulmonary circulation Some are single, many are multiple

Systemic arteriovenous aneurysms may be caused by trauma A large congenital arteriovenous aneurysm may cause high output cardiac failure in the neonatal period Small aneurysms are compatible with life for many years Such aneurysms in the brain are not rare and occasionally an aneurysm occurs in the myocardium between the coronary artery and the coronary vein The outstanding physical finding is a continuous murmur which is located over the area of the aneurysm

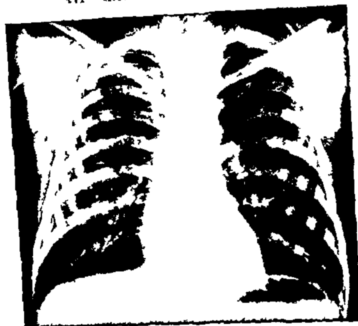


FIGURE XVI-3 Anomalous communication between the right pulmonary artery and the left auricle (Case XVI-2) Child

Treatment The patient was operated on by Dr Alfred Blalock on July 14 1948 The chest was entered on the right. A large soft, pulsating mass was found in the posterior part of the right lung The right pulmonary artery was exposed and a large thin walled vessel was found which arose from the posterior aspect of the right pulmonary artery and extended to the mass Further dissection showed that an anomalous vessel extended from the mass to the left auricle The anomalous vessel was ligated and divided The patient s color immediately improved No further surgery was attempted

Postoperative course The patient made an excellent recovery The pulsating mass in his forehead shrank until it became almost invisible One year and again three years after the operation the patient was re-examined Red blood cell count 4 6 million hemoglobin 15 gm hematocrit reading 39 oxygen saturation 93 per cent He remained well

Comment This patient had an anomalous vessel which arose from the right pulmonary artery and extended to a vascular mass which communicated directly with the left auricle This probably represented a developmental anomaly of the pulmonary vascular bed Functionally it was similar to an arteriovenous aneurysm Furthermore the condition was associated with a systemic arteriovenous aneurysm on his forehead

Treatment Cedilanid was given in the dispensary with but temporary improvement The only possible hope for the infant's life was ligation of the right external jugular artery and the carotid artery This operation was rapidly performed by Dr David Sabiston The bruit and thrill were entirely eliminated The heart rate immediately slowed to 60 beats per minute and continued to slow for the next half hour Thereafter the heart action ceased

Autopsy A partial autopsy was performed at the Lutheran Hospital A large vessel was found which connected the right external carotid artery, the middle meningeal artery, and the right lateral dural sinus

CASE XVI-2 *S X* (Harriet Lane Home, No A-63444, Johns Hopkins Hospital, No 467587) White male First seen at the Harriet Lane Home in 1948 at fifteen years of age * The chief complaint was abnormality of color

History Cyanosis was first noted at five years of age, when the patient was seen at another hospital At this examination he was found to have a pulsating lump on his forehead During the ensuing ten years the mass slowly increased in size and cyanosis deepened The patient s activity, however, was but slightly restricted Running caused dyspnea but he could walk a mile and had taken short Scout hikes

Physical examination Temperature 37 1°C , pulse 92 per minute respirations 20 per minute, blood pressure 120/80 mm of mercury

The patient was a small undernourished lad, intensely cyanotic with marked clubbing of the extremities and a small pulsating mass on his forehead The heart was normal in size The heart sounds were of good quality, no murmur was heard The remainder of the physical examination was negative

Laboratory data Red blood cell count 9 5 million hemoglobin 25 gm hematocrit reading 77

Electrocardiogram Normal sinus mechanism There was a slight right axis deviation but the unipolar precordial leads were normal

Teleoroentgenogram The contour of the heart was normal the vascular markings, however, were increased and showed minimal pulsations in the hilar shadows (see Figure XVI-3)

Angiocardiography Immediately after the dye entered the pulmonary artery a relatively large, rounded area was opacified in the right hilum and a fraction of a second later the dye was seen in the left auricle This finding suggested the presence of a large arteriovenous aneurysm in the right lung which poured blood directly into the left auricle (see Figure XVI-4)

Diagnosis Arteriovenous aneurysm in the right lung

* This case was also reported by Sloan and Cooley [8]

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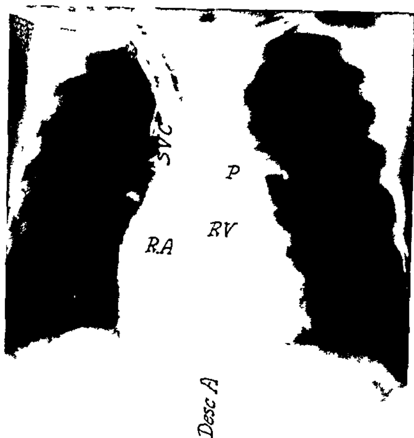




FIGURE XVI-4 Anomalous communication between the right pulmonary artery and the left auricle (Case XVI-2) Child

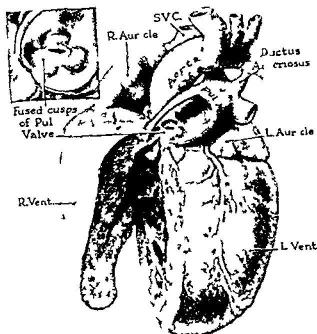


Figure XVII 1 Valvular pulmonary stenosis with an intact ventricular septum

poststenotic dilatation develops and eventually the pulmonary artery becomes greatly dilated The right ventricle is normally formed, the ventricular septum is intact

Infundibular stenosis is the name given to the stenosis which involves the infundibular portion of the right ventricle The malformation is caused by the failure in the expansion of the bulbus cordis[2] to form the outflow tract of the right ventricle in the normal manner The outflow tract of the right ventricle may be represented by a narrow pathway which leads to the pulmonary orifice or the infundibular chamber may be separated from the right ventricle by a muscular ridge in which there is only a small opening, as shown in Figure XVII 2 Although stenosis of the infundibular region is the rule in a tetralogy of Fallot its occurrence with an intact ventricular septum is the great exception

The narrowing of the pulmonary orifice whether it is valvular or infundibular renders it difficult for the right ventricle to pump the blood into the pulmonary artery Consequently the work of the right ventricle is greatly increased and the wall of the right ventricle becomes hypertrophied The smaller the opening into the pulmonary artery the greater is the difficulty in the ejection of

CHAPTER XVII

PULMONARY STENOSIS

OBSTRUCTION to the pulmonary blood flow is usually caused by an abnormality of the pulmonary valve, in some instances, however, the obstruction occurs within the right ventricle Both conditions are classified as "pure" pulmonary stenosis because the sole abnormality is the pulmonary stenosis, be it valvular or infundibular (see Section A) Occasionally constrictions occur in the branches of the pulmonary artery (see Section B) The last mentioned condition is designated as "peripheral" pulmonary stenosis

A *Pure Pulmonary Stenosis*

"Pure" pulmonary stenosis with an intact ventricular septum is not a rare malformation With the advent of cardiac catheterization and Sir Russel Brock s operation[1] for the alleviation of pulmonary stenosis, an extraordinary number of cases have been reported Furthermore, there are all grades of pulmonary stenosis

NATURE OF THE MALFORMATION

The stenosis is usually of the valvular type, in rare instances it may be located in the infundibulum of the right ventricle The degree of pulmonary stenosis is subject to great variation In some instances the pulmonary stenosis is so slight that the condition never causes symptoms and is compatible with a long and active life In other instances the pulmonary stenosis is so severe that the infant lives only a few months It has been the author's experience that the pulmonary orifice must be reduced to less than one third of its normal diameter for the condition to cause symptoms The opening may be only a millimeter or two in diameter In such instances the infant will die at an early age unless the stenosis is relieved by operation

The stenosis of the pulmonary valve results from a fusion of the three semilunar cusps over the pulmonary orifice to form a dome which has a central perforation, as shown in Figure XVII-1 Such is the location of the obstruction in approximately 95 per cent of the patients in whom pulmonary stenosis occurs with an intact ventricular septum

The pulmonary ring is almost always of normal size The pulmonary artery beyond the valve is usually normal at birth Over a period of years, however,

As the work of the right ventricle increases, the right ventricle fails to empty completely and the diastolic pressure in that chamber rises With the increase in the diastolic pressure, the right auricle encounters difficulty in emptying itself, consequently the work of the right auricle is increased The readily distensible right auricle undergoes both dilatation and hypertrophy

The valve covering the *foramen ovale* remains patent in approximately 75 per cent of patients with this malformation When this occurs, the high pressure in the right auricle will eventually push the valve away from the auricular septum and thereby cause functional patency of the foramen ovale Under such circumstances the foramen ovale acts as an escape valve for the relief of the high pressure in the right auricle and at the same time a right to-left shunt is established through it This in turn reduces the volume of blood which is directed into the right ventricle, consequently the right ventricular chamber remains relatively small but the wall of the right ventricle may be enormously thickened it may be more than 1 cm in thickness

COURSE OF THE CIRCULATION

During fetal life inasmuch as little blood flows to the lungs, even a severe degree of pulmonary stenosis places little strain upon the heart Although the increased pressure on the right side of the heart causes more than the usual quota of blood to flow through the foramen ovale to the left auricle and the left ventricle and thereby slightly increases the work of the left ventricle, the difference is not great At birth the heart is approximately normal in size

After birth the increased work demanded of the right ventricle by pulmonary stenosis increases the pressure in the right ventricle and, if this is great, it is transmitted back to the right auricle The increased pressure in that chamber tends to hold the foramen ovale open Therefore, if pulmonary stenosis is extreme the foramen ovale frequently remains widely patent

When the foramen ovale remains open, part of the blood from the right auricle flows into the right ventricle and some blood from the right auricle is shunted into the left auricle The blood which flows into the right ventricle is pumped out through the stenosed pulmonary orifice into the pulmonary artery, thence it flows slowly to the lungs The blood which reaches the lungs takes up its normal quota of oxygen The fully oxygenated blood is returned to the left auricle where it becomes mixed with the venous blood shunted from the right auricle into that chamber This mixture of venous and arterial blood flows from the left auricle into the left ventricle and thence is pumped out through the aorta

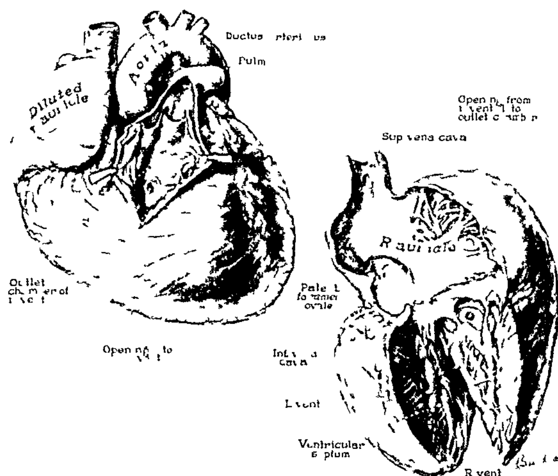


FIGURE XVII 2 Infundibular pulmonary stenosis with an intact ventricular septum (same patient as in Figure XVII-10) Adult

blood from the right ventricle, and the greater the work demanded of that chamber As the wall of the right ventricle hypertrophies, the musculature at the base of the pulmonary artery becomes thickened and eventually the muscular ring becomes so thick that it actually obstructs the outflow of blood from that chamber Consequently a vicious circle is set up which leads to still greater hypertrophy

Furthermore, it seems probable that the fibrotic valve grows more slowly than does the normal valve and therefore the stenosis of the pulmonary orifice becomes proportionally greater with the growth of the individual Certain it is that this malformation is one which may lead to progressive cardiac enlargement *Since there is no left to-right shunt, there is no increase in the volume of* blood which the right ventricle must pump Consequently there is little or no dilatation of the right ventricle The enlargement is almost entirely due to hypertrophy of the muscular wall

CLINICAL FINDINGS

The clinical findings[3,6] vary with the severity of the pulmonary stenosis and with the structure of the foramen ovale The more extreme the pulmonary stenosis, the greater is the tendency for the foramen ovale to remain patent Nevertheless, the foramen ovale may become completely sealed even though the pulmonary stenosis is extremely severe

If the pulmonary stenosis is slight, the patient is entirely asymptomatic It is only the presence of a harsh systolic murmur and a weak second sound which indicates that the murmur is of more than functional origin If, on the other hand the stenosis is severe, the condition causes progressive cardiac enlargement and, unless the obstruction is relieved, it eventually leads to cardiac failure

The appearance of the patient is remarkable only in that he is usually strong and well developed Infants are often exceptionally sturdy, growth and development are normal

Dyspnea is the outstanding complaint The dyspnea is related to the patient s inability to increase the pulmonary blood flow rather than to a venous-arterial shunt If the pulmonary stenosis is extreme, the patient simply cannot appreciably increase the pulmonary blood flow with exercise This means that he is unable to increase the minute output of the heart and consequently cannot increase the supply of oxygen to the body or to the brain Therefore, even though the patient shows no cyanosis and he has a normal arterial oxygen saturation, he becomes dyspneic on slight exertion and stops to rest Infants with this type of pulmonary stenosis rarely suffer from attacks of paroxysmal dyspnea, children seldom squat when tired

It is important to remember that complaints are late manifestations of this malformation Many patients are asymptomatic until early adult life This fact is vividly illustrated by two patients whom the author has seen One was a man who had served in the Merchant Marine during World War II and at twenty two years of age developed severe cardiac failure The other had been a top athlete in his college class but at twenty four years of age was so incapacitated that he could scarcely walk across the room Indeed, because of his history of athletic prowess and his strong physique combined with the fact that he spent most of the time sitting indolently, one of the author s assistants raised the question whether he could be malingering The enormous cardiac enlargement and chronic cardiac failure clearly indicated that his complaints were real Two days later he keeled over in bed and was dead Fortunately, with the advent of cardiac surgery, this sequence of events is seldom, if ever, seen

to the systemic circulation and is returned in the normal manner by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again (see Diagram xvii–1) It is the establishment of a right-to-left shunt through the foramen ovale which renders the patient cyanotic

Since the venous arterial shunt in this malformation is established through the foreman ovale, as long as the foramen ovale is functionally closed and in all instances in which the foramen ovale becomes completely sealed, the course of the circulation is normal The blood from the right auricle flows into the right ventricle and is pumped out with difficulty through the pulmonary orifice into the pulmonary artery Inasmuch as the pulmonary pressure is low, the blood in the pulmonary artery flows slowly to the lungs Nevertheless, all the blood which goes to the lungs is fully oxygenated and is returned in the normal fashion by the pulmonary veins to the left auricle and the left ventricle From the left ventricle the blood is pumped out by way of the aorta to the systemic circulation and is returned by the superior and inferior venae cavae to the right auricle, there the cycle starts again, as shown in Diagram xvii–2

PHYSIOLOGY OF THE MALFORMATION

The pulmonary stenosis causes difficulty in the expulsion of blood from the right ventricle but it also protects the lungs from the high pressure generated in that chamber Consequently the difficulty imposed upon the circulation is primarily that of the increased work required of the right ventricle If the pulmonary stenosis is extreme, the pulmonary blood flow is low and the systemic blood flow is correspondingly low Nevertheless, if the foramen ovale is sealed, the oxygen saturation of the arterial blood is normal The slow circulation enables the body to take up a large amount of oxygen and nutrition from the blood as it passes through the capillaries The nutrition of the individual is correspondingly good

The stenosis of the pulmonary valve not only increases the work required of the right ventricle to pump the blood into the pulmonary artery, but it also breaks the force with which the blood is ejected into the pulmonary artery, and, furthermore, that force is dissipated in all directions Blood pools in the main pulmonary artery and flows slowly to the lungs The dissipation of the energy generated in the right ventricle is the most probable explanation of the post stenotic dilatation of the pulmonary artery Be that as it may, poststenotic dilatation of the pulmonary artery is the rule In most instances the longer the duration of the pulmonary stenosis and the greater its severity, the greater is the post stenotic dilatation of the pulmonary artery

DIAGRAM XVII-1

Valvular pulmonary stenosis with an intact ventricular septum and a patent foramen ovale

The essential feature of this malformation is the stenosis of the pulmonary valve combined with patency of the foramen ovale The ductus arteriosus is completely obliterated

The blood from the right auricle flows both into the right ventricle and through the foramen ovale into the left auricle. The blood from the right ventricle is forced through the stenosed pulmonary orifice to the lungs where it is oxygenated, and the oxygenated blood is returned by the pulmonary veins to the left auricle, where it meets the venous blood which has been forced from the right auricle through the foramen ovale into the left auricle the admixture of venous and arterial blood flows into the left [illegible] by way of the aorta to the systemic circulation and is returned by the

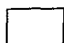
[illegible]

and hypertrophy of that chamber The increased pressure in the right ventricle is transmitted back into the right auricle which becomes enlarged The patency of the foramen ovale acts as an escape valve consequently there is less hypertrophy and dilatation of the right auricle under these conditions than when the foramen ovale is completely closed Moreover there is less engorgement of the liver and it seldom pulsates

Inasmuch as there is high pressure on the right side of the heart and patency of the foramen ovale a right-to-left shunt is established between the two auricles The extent of the oxygen unsaturation of the arterial blood depends upon the volume of venous blood shunted from the right auricle to the left auricle As this increases the patient develops cyanosis and polycythemia

Clinical diagnosis The intensity of the cyanosis varies with the severity of the pulmonary stenosis and the extent of patency of the foramen ovale When the stenosis is extreme cyanosis may date from birth Usually however cyanosis appears between the second and the sixth year of life and slowly increases in intensity Dyspnea is more marked than cyanosis Examination of the heart shows a basal *systolic murmur* and a thrill The second sound at the base is weak or absent There is progressive enlargement of the right ventricle and the right auricle There is fullness of the pulmonary conus which also becomes progressively more marked because of the poststenotic dilatation of the pulmonary artery The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy Terminally the heart becomes enormously enlarged and there is engorgement of the liver and there may be both edema and ascites The condition should however be corrected by surgery before the heart becomes greatly enlarged

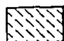
Diagram XVII-1



Arterial blood (fully saturated)



Venous and arterial blood
Cyanosis visible



Small admixture of venous blood
No visible cyanosis



Venous blood

DIAGRAM XVII–2

Valvular pulmonary stenosis with an intact ventricular septum

The sole abnormality in this malformation is the stenosis at the orifice of the pulmonary artery. The three semilunar valves are fused together to form a dome with a central perforation. The ductus arteriosus is completely obliterated, the foramen ovale is closed.

The blood from the right auricle flows into the right ventricle and that which can be forced through the pulmonary orifice goes to the lungs and is returned by the pulmonary veins to the left auricle. Thence it flows to the left ventricle and out by way of the aorta to the systemic circulation and is returned by the superior and inferior venae cavae to the right auricle.

The obstruction to the outflow of blood from the right ventricle causes dilatation and hypertrophy of that chamber. The increased work required of the right ventricle raises the pressure in that chamber and this in turn increases the work required of the right auricle; it, too, is dilated and hypertrophied. The inferior vena cava is dilated and the liver is engorged and pulsates.

Inasmuch as there is no abnormal communication between the two circulations there is no venous-arterial shunt. Consequently the oxygen saturation of the arterial blood is normal.

Clinical diagnosis. Cyanosis is absent. Dyspnea may be extreme. There is a basal systolic murmur and a thrill. The second sound at the base is weak or absent. The heart undergoes progressive enlargement. The enlargement is mainly due to hypertrophy of the right ventricle and to dilatation and hypertrophy of the right auricle. There is fullness of the pulmonary conus due to poststenotic dilatation of the main pulmonary artery. The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy. Terminally the liver is engorged and pulsates and there may be both edema of the extremities and ascites.

The condition can and should be corrected by surgery as soon as there is evidence of cardiac strain.

DIAGRAM XVII–2

Head and upper extremities

R Lung

L Lung

P.A.

S.V.C.

Aorta

Pul. A.

P.V.

Pulmonary Artery

Pul V

L.A.

Descending

R.A.

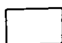
L.V

R.V

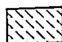
I.V.C.

Aorta

Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

are more frequently palpable with a closed auricular septum or with slight patency of the foramen ovale than when the valve is widely patent Regardless of the presence or absence of cyanosis, pulsations of the liver are indicative of high pressure in the right auricle and hence of a severe degree of pulmonary stenosis Therefore this finding is a strong indication for prompt operation

Edema and ascites may occur as late manifestations of the right sided heart failure Nevertheless, owing to the difficulty in the expulsion of blood from the right ventricle, pulmonary congestion is rare

Episodes of collapse and loss of consciousness are not uncommon terminal manifestations, because the patient suffers from a low pulmonary blood flow and a correspondingly low systemic blood flow Indeed, there may be a striking discrepancy between the patient s sturdy growth and his history of former activity and the sudden development of lassitude and exhaustion As a matter of fact the pulmonary blood flow may become so meager that life just peters out

CARDIAC FINDINGS

The size of the heart and the rate of enlargement vary with the severity of the pulmonary stenosis If the pulmonary stenosis is slight to moderate, the malformation does not cause cardiac enlargement When the pulmonary stenosis is severe, the condition leads to progressive enlargement *Cardiac enlargement precedes symptoms* Therefore it is extremely important for patients suspected of this malformation to be kept under close observation This is especially true for infants as cardiac enlargement may occur with extraordinary rapidity Moreover, months are to an infant as years are to a child of ten Therefore the infant should be followed at monthly intervals for evidence of increase in heart size The two x rays in Figure XVII–3 show a tremendous increase in the size of the heart This occurred in an infant between six months and one year of age If the heart becomes definitely enlarged or the infant develops pulsations at the margin of the liver or shows signs of cardiac failure, prompt operation is indicated

In children however cardiac enlargement occurs more slowly For this reason changes in the electrocardiogram are a more sensitive index to increasing cardiac strain than are the changes in the x ray (see Electrocardiographic Findings) Cardiac enlargement may also occur rapidly during the spurt of growth which occurs at puberty Therefore, at this age, changes in the size of heart again become significant

The rate of enlargement depends upon the amount of work required of the

The occurrence of cyanosis depends primarily upon the structure of the auricular septum[4] If the auricular septum is intact and the foramen ovale is completely sealed, there is no communication between the two sides of the heart Even though the pressure in the right auricle is greatly increased, there is no venous arterial shunt Cyanosis is absent The oxygen saturation of the arterial blood is normal

Since the *foramen ovale is normally patent at birth, in the presence of pulmonary stenosis* a right to-left shunt may be readily established immediately after birth For this reason infants with pulmonary stenosis frequently show cyanosis at birth Generally, however, the foramen ovale tends to close, the right to-left shunt lessens, and not infrequently cyanosis entirely disappears

On the other hand, if the foramen ovale remains patent, as the pulmonary stenosis increases the pressure in the right ventricle rises and this in turn increases the pressure in the right auricle, under such circumstances the foramen ovale is again forced open and the right to-left shunt is established When the volume of the shunt is sufficiently great, the patient develops cyanosis Consequently, although cyanosis due to patency of the foramen ovale may date from birth, or may be present at birth and subsequently disappear, it frequently develops insidiously and becomes apparent when the patient is between two and seven years of age Thereafter, unless the stenosis is relieved, the cyanosis gradually increases in intensity

Polycythemia develops as the oxygen unsaturation of the arterial blood increases The red blood cell count, the amount of available hemoglobin, and the hematocrit reading may reach as high levels in these patients as in any other patient who has a persistent venous arterial shunt The increase in the hemoglobin in turn increases the intensity of the cyanosis

Clubbing of the extremities gradually develops as the polycythemia increases

The venous pressure becomes increased when the pressure in the right auricle becomes abnormally high

The neck vessels become engorged and often show conspicuous pulsations

The liver gradually enlarges and frequently extends from two to three finger breadths below the costal margin Owing to the high pressure on the right side of the heart and the difficulty in the expulsion of blood from the right auricle, the forceful contractions of the right auricle are frequently transmitted back to the margin of the liver Such pulsations are presystolic in time and may occur with minimal engorgement of the liver Inasmuch as the foramen ovale acts as an escape valve for the high pressure in the right auricle, pulsations in the liver

are more frequently palpable with a closed auricular septum or with slight patency of the foramen ovale than when the valve is widely patent Regardless of the presence or absence of cyanosis, pulsations of the liver are indicative of high pressure in the right auricle and hence of a severe degree of pulmonary stenosis Therefore this finding is a strong indication for prompt operation

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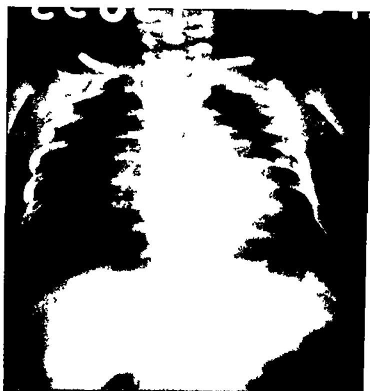
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The rate of enlargement depends upon the amount of work required of the



At six months



At one year

FIGURE XVII–3 Severe valvular pulmonary stenosis with an intact ventricular septum Infant

right ventricle Therefore the size of the heart varies not only with the severity of the pulmonary stenosis, but also with the load placed on the heart by physical exertion The author has seen one woman with severe pulmonary stenosis and a pressure of nearly 200 mm of mercury in the right ventricle who had lead such a quiet, *restricted* life that her heart had remained normal in size Usually severe pulmonary stenosis leads to great cardiac enlargement

Rapid increase in the size of the heart during childhood at the period when growth is slow should suggest the *possibility that the stenosis is infundibular, not* valvular, in type

A harsh systolic murmur and a thrill are produced as the blood is forced through the stenosed pulmonary orifice into the pulmonary artery Both the murmur and the thrill are maximal over the pulmonary area The thrill is often readily palpable in the suprasternal fossa In cases of severe pulmonary stenosis and great cardiac enlargement, the murmur is well heard high up beneath the left clavicle and is frequently well transmitted to the interscapular region The fibrosis of the valve, combined with the low pressure in the pulmonary artery, prevents an abrupt or snapping closure of this valve, consequently the pulmonic s cond sound is weak or absent. Nevertheless, the murmur ends abruptly with the end of systole The pulmonary valve is usually competent, diastole is clear It is the harsh systolic murmur that ends abruptly, combined with a weak or absent second sound, which gives the clue to the diagnosis

It is important to remember that an infant may have such an extreme degree of pulmonary stenosis that there *is no murmur or thrill, such an infant suffers* from rapid cardiac enlargement and cardiac failure associated with an engorged pulsating liver

In the presence of great cardiac enlargement there is usually an apical systolic murmur which is transmitted to the axilla and also a gallop rhythm

The characteristic thrill in the suprasternal notch is absent when there is in fundibular stenosis even when the murmur and thrill are maximal high up in the second interspace to the left of the *sternum* The murmur and the thrill are frequently of maximal intensity at a lower level in infundibular stenosis than in valvular stenosis of a comparable degree Occasionally the second sound is audible over the base of the heart, this *finding* is also suggestive of an infundibular stenosis

Cardiac failure may occur in the neonatal period without great cardiac enlargement It *is manifested* by cyanosis and by engorgement of the liver with pulsations at its margin There may even be *generalized edema* The infant may

be greatly helped by digitalis As the baby regains compensation, cyanosis lessens and may disappear and thereafter he may do well for a number of months or even for years

In contrast to this, if the infant does well in the neonatal period but during the subsequent months develops cardiac enlargement and cardiac failure, early operation is urgently indicated (see under Treatment)

In children and adults cardiac failure is a late manifestation As long as the heart is able to enlarge to meet the needs of the individual, the patient remains asymptomatic Consequently, by the time cardiac failure develops, the heart is always greatly enlarged The liver is engorged and usually pulsations are palpable at its margin Edema develops and subsequently there may be ascites The lungs remain relatively clear Even at this time prompt relief of the pulmonary stenosis may be of great benefit to the patient

X RAY AND FLUOROSCOPIC FINDINGS

The x ray and fluoroscopic findings are distinctive The second curve to the left of the sternum is exaggerated If the stenosis is slight or moderate, the heart may be normal in size Usually the right ventricle is hypertrophied and the heart appears to be slightly to moderately enlarged Owing to the development of poststenotic dilatation of the pulmonary artery, in children and adults the main pulmonary artery is usually enlarged and there is fullness in the region of the pulmonary conus, as shown in Figures XVII–4, 5, 6, and 15

When the pulmonary stenosis is extreme, the heart may become enormously enlarged The enlargement of the right ventricle is so great that it not only presses against the anterior chest wall but also displaces the left ventricle backward and the pulmonary artery upward The greatly dilated pulmonary artery lies above the thick walled right ventricle and the pulmonary valve and the main segment of the pulmonary artery lie at an abnormally high level

The absence of poststenotic dilatation of the pulmonary artery is sometimes considered indicative of infundibular stenosis There are, however, many exceptions The characteristic fullness of the pulmonary conus is frequently absent in early infancy, as poststenotic dilatation takes time to develop Indeed, in children with extreme pulmonary stenosis (see Figure XVII–8), so little blood may be pumped through the pulmonary orifice that poststenotic dilatation of the pulmonary artery does not occur Furthermore, the characteristic fullness of the pulmonary conus may occur in children with infundibular stenosis as illustrated in Figure XVII–9 Nevertheless, in older patients with great cardiac enlargement, the

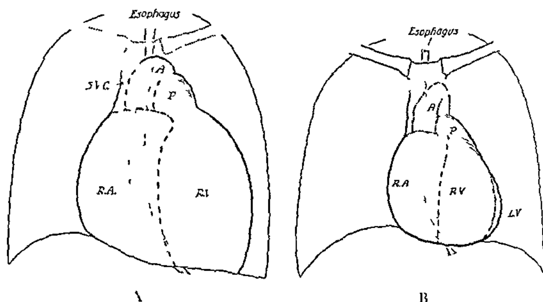


Anterior posterior position



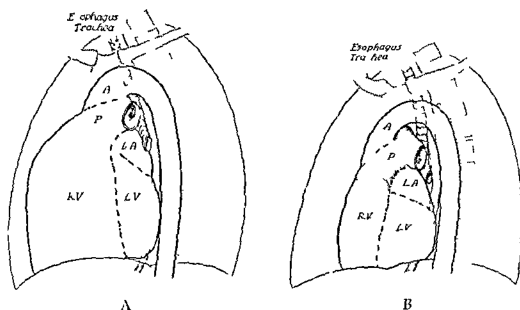
Left anterior oblique position

FIGURE XVII-4 Valvular pulmonary stenosis with an intact ventricular septum (same patient as in Figures XVII-13 14) Child



A B

ANTERIOR POSTERIOR POSITION



A B

LEFT ANTERIOR OBLIQUE POSITION

FIGURE XVII-5 (A) Valvular pulmonary stenosis with an intact ventricular septum and (B) normal heart Child



FIGURE XVII-6 Valvular pulmonary stenosis with an intact ventricular septum (same patient as in Figure XVII-7) Adult

absence of fullness of the pulmonary conus, as shown in Figure XVII-10, usually is indicative of infundibular stenosis

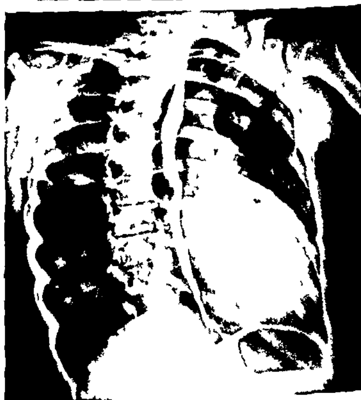
In the left anterior-oblique position the right ventricle is usually seen to be enlarged (see Figures XVII-4, 5, and 7) When the heart is greatly enlarged, the right ventricle becomes flattened against the sternum[4] and further enlargement of the right ventricle displaces the left ventricle backward so that the heart does not clear the spinal column until the patient is rotated to an angle of 60 degrees Under such circumstances, it is usually possible to detect the interventricular groove The posterior position of this groove adds confirmatory evidence that the increase in the size of the heart is mainly due to the great right ventricular hypertrophy (see Figure XVII-5)

Examination in the right anterior-oblique position contributes little, except that the left auricle is of normal size and the esophagram is normal

Fluoroscopic examination aids still further in the diagnosis as there is a striking discrepancy between the size of the pulmonary conus and the pulsations



Left anterior-oblique position



Right anterior oblique position

FIGURE XVII-7 Valvular pulmonary stenosis with an intact ventricular septum (same patient as in Figure XVII-6) Adult



Before operation



After operation

FIGURE XVII-8 Severe valvular pulmonary stenosis with an intact ventricular septum Child



FIGURE XVII-9 Infundibular pulmonary stenosis with an intact ventricular septum Child



FIGURE XVII-10 Infundibular pulmonary stenosis with an intact ventricular septum (same patient as in Figure XVII-2) Adult

in the main branches of the pulmonary arteries Even if there are visible pulsations in the pulmonary conus, the pulsations in the left and right branches of the pulmonary artery are minimal or absent The greater the cardiac enlargement, the quieter are the hilar shadows In patients with minimal cardiac enlargement, a hilar dance may be discerned, in those with patency of the foramen ovale there is an actual reduction in the pulmonary blood flow and the lungs are proportionately clear

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram is of great aid not only in the diagnosis of this malformation but in an estimation of the severity of the pulmonary stenosis A right axis deviation and evidence of right ventricular hypertrophy are characteristic of pure pulmonary stenosis Furthermore, as the work of the right ventricle increases, the unipolar precordial leads show evidence of progressively greater right ventricular hypertrophy and so-called right ventricular strain'

When the pulmonary stenosis is extremely mild, the electrocardiogram may show only a right axis deviation and the unipolar precordial leads will be normal As the work of the right ventricle increases, the unipolar precordial leads reflect the progressive right ventricular hypertrophy When the pressure is under 80 mm of mercury, the R wave in V_1 is usually prominent but the T wave remains upright As the pressure in the right ventricle increases, the T waves in V_1 become inverted This usually does not occur until the pressure is between 100 and 120 mm of mercury As the pressure in the right ventricle rises still further, the pattern of so-called right ventricular strain develops that is, a slight delay in the onset of the intrinsicoid deflection of V_1 and a tall R wave combined with inversion of the T waves in V_1, possibly in V_3 and even in V_5, the T wave in V_6 tends to become upright the S waves over the left precordium are deep Such changes (see Figure XVII–11) are usually indicative of a right ventricular pressure of more than 140 mm of mercury[2]

The changes in the electrocardiogram usually become apparent before there is any detectable enlargement in the x ray Therefore serial electrocardiograms are extremely useful in the evaluation of the condition of the patient Careful evaluation of the electrocardiographic findings eliminates the necessity for cardiac catheterization

In childhood electrocardiographic evidence of rapidly progressive right ventricular strain which is out of proportion to an increase in the size of the heart is always suggestive that the stenosis is infundibular and not valvular

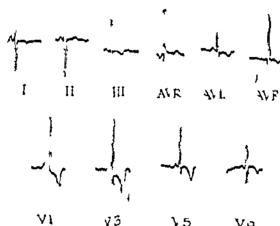


FIGURE XVII-11 Valvular pulmonary stenosis

The P waves reflect the amount of work required of the auricle As the pressure in the right ventricle rises, the pressure in the right auricle also rises and the right auricle undergoes dilatation and hypertrophy Under such circumstances the P waves are both broad and tall, consequently they resemble the Appalachian Mountains rather than the tall, sharp peaks of the Himalayas

SPECIAL TESTS

The circulation time (arm to tongue) is usually normal or prolonged Even though the foramen ovale is widely patent, sufficient test material is seldom shunted through the foramen ovale to the systemic circulation to give a short circulation time The circulation time is commonly from twelve to fifteen seconds in a child and may be over twenty seconds in an adult In infants the normal circulation time is so short that the test is seldom significant indeed, in the presence of a widely patent foramen ovale the circulation time may be phenomenally short

The oxygen saturation of the arterial blood varies with the amount of venous blood which is shunted through the foramen ovale If the foramen ovale is widely patent, the oxygen saturation of the arterial blood may be greatly reduced Usually, however, even in the presence of persistent cyanosis, the arterial oxygen saturation is relatively high and falls but slightly with exercise In patients with an intact auricular septum, the oxygen saturation of the arterial blood is normal

The exercise test usually shows no increase in the oxygen consumption per liter of ventilation [8] In other words, the pulmonary stenosis prevents the increase in the pulmonary blood flow which normally occurs during exercise

Cardiac catheterization reveals a high pressure in the right ventricle and a

low pressure in the pulmonary artery, frequently the pressure in the right auricle is also increased The oxygen content of the blood in the right auricle, in the right ventricle, and in the pulmonary artery should be essentially the same Although the over all shunt is from right to left, if the catheter is drawn into the stream of the shunt and the sample is taken close to the opening in the auricular septum the oxygen content of this sample may be higher than that taken from the superior vena cava, thereby giving the impression that there is a gross defect in the auricular septum Occasionally, even though there is no gross defect in the auricular septum, the catheter may be passed through the foramen ovale into the left auricle When the pulmonary stenosis is extreme, it may be difficult or impossible to pass the catheter into the pulmonary artery

If the pressure in the right ventricle exceeds systemic pressure, it is strong presumptive evidence that the ventricular septum is intact It is, however, essential to catheterize the pulmonary artery because only the combination of high pressure in the right ventricle and low pressure in the pulmonary artery proves the existence of pulmonary stenosis Figure XVII–12 shows the abrupt change in the pressure at the level of the pulmonary valve as the catheter is withdrawn from the pulmonary artery into the right ventricle

A few words of caution in regard to catheterization are necessary Serious symptoms may develop if the size of the catheter is such as to occlude the pulmonary orifice and thereby cut off the supply of blood to the lung Furthermore, in the presence of great right ventricular hypertrophy there is real danger that catheterization of the right ventricle may precipitate ventricular fibrillation Finally it should be remembered that with myocardial failure, the pressure in the right ventricle will fall

Cardiac catheterization may be of value in the differentiation of infundibular from valvular stenosis In valvular stenosis the catheter passes directly from an area of high pressure to an area of low pressure When the stenosis lies within the ventricle, the catheter passes from the area of high pressure through an area of lower ventricular pressure before it enters the pulmonary artery Furthermore, when one is unable to pass the catheter into the pulmonary artery, the obstruction is usually encountered at a lower level in an infundibular stenosis than in a stenosis of the valvular type Nevertheless, unless the physician is on his guard, the differences upon catheterization may be overlooked

Angiocardiography shows prompt opacification of the right side of the heart, first the right auricle and then the right ventricle is filled It is usually possible to demonstrate that the right ventricle is thick walled as the dye does not extend

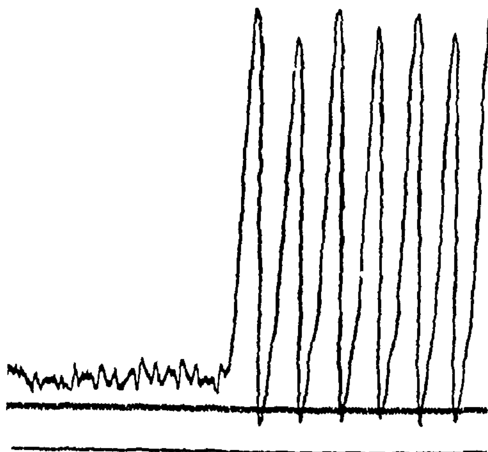


FIGURE XVII–12 Valvular pulmonary stenosis

Pressure tracing showing abrupt change in pressure between the pulmonary artery and the right ventricle Pressure recording pulmonary artery 22/9 mm Hg right ventricle 140/4 mm Hg

to the margin of the cardiac silhouette The pulmonary artery is promptly opacified Nevertheless, owing to the pulmonary stenosis, only a small amount of dye is ejected into the pulmonary artery with each ventricular systole Consequently, although the main pulmonary artery fills immediately, it continues to fill over a long period of time The aorta is not promptly delineated and is never opacified before the left auricle and the left ventricle are filled The latter are always better visualized in the lateral position than in the anterior posterior position Therefore angiocardiograms in both positions are important When there is patency of the foramen ovale due to the constant shunting of a small amount of blood through the foramen ovale, the aorta is usually faintly outlined and dye in small concentration remains in the aorta for several seconds and then disappears Inas

much as the blood flows slowly from the pulmonary artery to the lungs, dye in high concentration may remain in the main *pulmonary* artery after it has disappeared from the aorta Figures XVII–13 and 14 *show the lingering of the dye in* the pulmonary artery in a child with 'pure' pulmonary stenosis

In contrast to this, the angiocardiogram of an infant with a widely patent foramen ovale may show *such prompt* and dense opacification of the aorta simultaneously with the delineation of the pulmonary artery that the *condition may* be mistaken for a tetralogy of Fallot

A word of warning in regard to angiocardiography if the pulmonary stenosis is *extreme, throughout* the time that the dye is being ejected into the pulmonary artery, the circulation of blood to the lungs is virtually cut off *For this* reason angiocardiography is dangerous for patients with severe pulmonary stenosis, it is especially dangerous for those with an extremely low arterial oxygen saturation Indeed, the deprivation of oxygen may be fatal

DIAGNOSIS

The outstanding features of this malformation are a harsh systolic murmur and a thrill over the pulmonary area, combined with a weak or absent second sound at the base The patient may or may not show cyanosis The insidious development of cyanosis in childhood is common The demonstration of pulsations at the margin of the liver indicates that the stenosis is extremely severe

The electrocardiogram shows a right axis deviation and generally shows the pattern of right ventricular hypertrophy X ray examination almost invariably shows fullness of the pulmonary conus and enlargement of the main branches of the pulmonary artery Upon fluoroscopy there is a discrepancy between the prominence of the pulmonary conus and the pulsations in the hilar shadows the former is conspicuous the latter are minimal or absent

The diagnosis can be definitively established by cardiac catheterization provided the pulmonary artery is catheterized When there is no evidence of a left to-right intracardiac shunt the finding of high pressure in the right ventricle combined with low pressure in the pulmonary artery is indicative of pulmonary stenosis with an intact ventricular septum

The differentiation of infundibular from valvular stenosis although difficult and indeed sometimes impossible, is extremely *important In children* the evidence of rapid cardiac enlargement in the x ray or of rapidly progressive right ventricular hypertrophy in the electrocardiogram should suggest that the stenosis is infundibular The finding of a murmur maximal in the second left inter

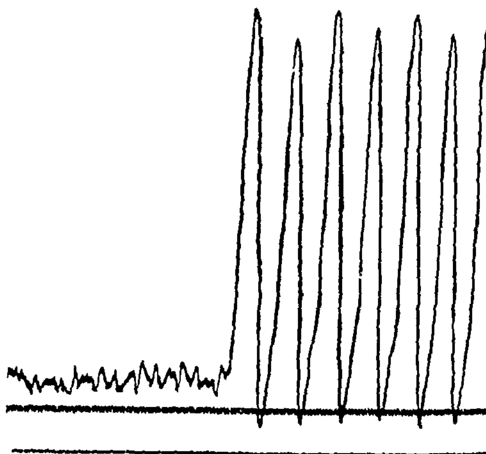


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At four seconds



At seven seconds

FIGURE XVII-14 Valvular pulmonary stenosis with an intact ventricular septum (series from Figure XVII-13 continued same patient as in Figure XVII-4) Child

Note dye still visible in the main pulmonary artery at seven seconds



At one second



At two seconds

FIGURE XVII-15 Valvular pulmonary stenosis with an intact ventricular septum (same patient as in Figure XVII-4) Child

Ebstein's anomaly of the tricuspid valve when extreme, may closely resemble severe long standing pulmonary stenosis In Ebstein's anomaly fatigue is the outstanding complaint, cyanosis is more marked than dyspnea, the heart sounds are weak, the murmurs are blurred and confused, and arrhythmias are common Compensation is frequently precarious but pulsations at the margin of the liver are usually absent The electrocardiogram usually differentiates the two conditions in Ebstein's anomaly the deflections in V_1 are of low amplitude and the QRS is of long duration, whereas in pulmonary stenosis the R wave in V_1 is tall Angiocardiography shows enormous dilatation of the right auricle and an extremely thin walled right ventricle Cardiac catheterization shows increased pressure in the right auricle and normal pressure in the right ventricle and in the pulmonary artery

Complete transposition of the great vessels may be confused with pulmonary stenosis during the first weeks of life In both conditions cyanosis may date from birth the heart is normal in size, there is no fullness of the pulmonary conus, and the vascular markings in the lungs are normal or slightly increased Moreover, both conditions may cause cardiac failure at an early age In pure pulmonary stenosis however, the electrocardiogram shows greater evidence of right ventricular hypertrophy than is usual in a complete transposition of the great vessels The clinical course is also different In pure pulmonary stenosis the infant usually responds to digitalis and regains compensation cyanosis gradually diminishes and may entirely disappear Thereafter the infant gains weight and does well In a complete transposition of the great vessels of such a type as to cause cyanosis and cardiac failure in the neonatal period, although the infant may be temporarily helped by digitalis the clinical course is one of increasing cyanosis rapid cardiac enlargement and cardiac failure Death usually occurs within the first few weeks of life

A tetralogy of Fallot may be confused with an isolated valvular pulmonary stenosis in early infancy In a tetralogy of Fallot the lung fields are excessively clear Furthermore the heart is usually normal in size, whereas a severe degree of pulmonary stenosis with an intact ventricular septum leads to great cardiac enlargement A child with a tetralogy of Fallot develops cyanosis at an early age and usually squats when tired the heart is small, the pulmonary artery is inconspicuous the liver does not pulsate and the circulation time is short The electrocardiogram shows right axis deviation and evidence of right ventricular hypertrophy but usually does not show the pattern of extreme right ventricular hypertrophy In doubtful cases cardiac catheterization may be necessary to differ

space and no thrill in the suprasternal notch is also strongly suggestive of infundibular stenosis *In adults* the absence of demonstrable poststenotic dilatation of the pulmonary artery or the low level at which the systolic murmur is maximal is suggestive of an infundibular stenosis

DIFFERENTIAL DIAGNOSIS

In the absence of cyanosis "pure" pulmonary stenosis may be confused with an auricular septal defect or with pulmonary hypertension Both the non-cyanotic and the cyanotic forms may be mistaken for an Eisenmenger complex and occasionally may be confused with Ebstein's anomaly of the tricuspid valve In early infancy the cyanotic form may be mistaken for a complete transposition of the great vessels, both in infancy and in childhood the condition may occasionally be mistaken for a tetralogy of Fallot and for defective development of the right ventricle with pulmonary stenosis

An auricular septal defect is differentiated from "pure" pulmonary stenosis by a slightly accentuated second sound at the base of the heart, which is usually reduplicated Furthermore, fluoroscopy reveals the presence of a hilar dance The electrocardiogram usually shows a right bundle branch block Furthermore, the patient has a frail build but does not suffer from severe dyspnea In pure pulmonary stenosis the second heart sound is weak or absent and, although the pulmonary artery may be conspicuous, the pulsations in its branches are minimal The unipolar precordial leads show evidence of right ventricular hypertrophy The patient, although of sturdy stature, may become dyspneic on slight exertion

Pulmonary hypertension is differentiated by the fact that the pulmonic second sound is markedly accentuated and is usually reduplicated Indeed, it is only in the presence of myocardial failure that the second sound may become so weak that the two conditions are confused Furthermore, pulmonary insufficiency is relatively common with pulmonary hypertension and rarely occurs with pulmonary stenosis

The Eisenmenger complex is differentiated from "pure" pulmonary stenosis by the absence of cyanosis and dyspnea in infancy and early childhood, by the presence of a hilar dance, and by the fact that the circulation time is short In older patients angiocardiography will show clear evidence of an over riding aorta Cardiac catheterization will show that the oxygen saturation in the pulmonary artery is higher than that in the right ventricle and that the pressure in the pulmonary artery is abnormally high

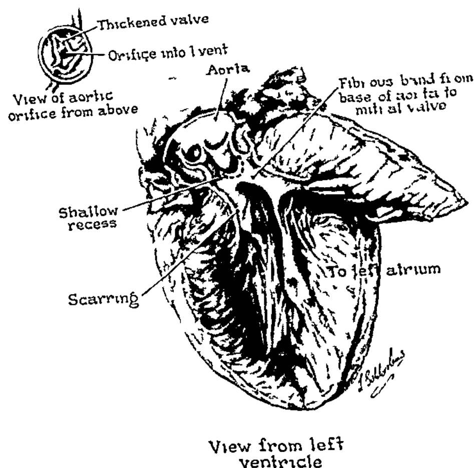


FIGURE XVII 16 Valvular pulmonary stenosis combined with subvalvular aortic stenosis (same patient as in Figures XVII 17 18) Child

valvular pulmonary stenosis an intact ventricular septum and a gross defect in the auricular septum

Pulmonary stenosis may occur in combination with aortic stenosis [9, 1] The author has seen one such instance in a five year old child who proved to have a valvular pulmonary stenosis and subvalvular aortic stenosis (see Figure XVII-16) This case was reported by Sissman et al [11] The patient had a harsh systolic murmur maximal over the base of the heart, which obliterated the heart sounds The second sound was inaudible to the right as well as to the left of the sternum The child suffered from progressive cardiac enlargement and cardiac failure The



FIGURE XVII–15 Valvular pulmonary stenosis an auricular septal defect, and an intact ventricular septum Adult

entiate the two conditions If the right ventricular pressure is above systemic pressure, it is strong presumptive evidence that the ventricular septum is intact

Defective development of the right ventricle with pulmonary stenosis differs from 'pure pulmonary stenosis in that the right ventricle is a small chamber which is unable to carry its full load Because of the decreased pulmonary blood flow and the small size of the pulmonary artery, the shadow at the base of the heart to the left of the sternum is concave In the left anterior oblique position the x ray shows that the left ventricle as well as the right ventricle is enlarged The electrocardiogram shows evidence of right ventricular hypertrophy in V_1 and left ventricular dominance in V and V_6 Angiocardiography shows that the right ventricle is a tiny chamber

COMMONLY ASSOCIATED ANOMALIES

A gross defect in the auricular septum of the ostium secundum type may occur in combination with valvular pulmonary stenosis Under such circumstances the defect in the auricular septum acts as an escape valve Consequently the more severe the pulmonary stenosis, the greater is the right to left shunt and the deeper is the cyanosis The patient may be severely limited but the heart remains remarkably small Figure XVII–15 illustrates the case of an adult with a severe

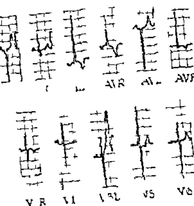


FIGURE XVII-18 Valvular pulmonary stenosis combined with subvalvular aortic stenosis (same patient as in Figure XVII-16) Child

Prophylactic antibiotics are indicated throughout life for the prevention of subacute bacterial endocarditis when there is danger of infection from dental extractions or other procedures Even after successful operation, the pulmonary valve is not normal the patient is still susceptible to the disease

Limitation of exercise is rarely necessary, as the condition can be relieved by surgery Therefore only if surgical treatment is unavailable is restriction of activity indicated In most parts of the world a child with a valvular pulmonary stenosis may be permitted to lead a normal life, if his activity places a strain on his heart, operation is indicated

The advisability of surgical treatment depends upon the severity of the stenosis In some instances the pulmonary stenosis is so slight that the heart is normal in size, the pulmonary blood flow can be increased with exercise, and the electrocardiogram does not even show evidence of right ventricular hypertrophy Such a patient is asymptomatic and if he has attained his full growth, he will certainly remain so The condition need cause no concern The prognosis is excellent without operation

In the author s opinion, if the heart remains normal in size and the patient is asymptomatic and able to lead a normal life and the pressure is under 100 mm of mercury in the right ventricle, operation is *not indicated*, even if the electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy It must be remembered that operation does *not* restore the valve to normal and that mild pulmonary stenosis is compatible with a long and active life

Surgical treatment is indicated if there is evidence of *severe* or *progressive* cardiac strain Inasmuch as cardiac enlargement precedes symptoms, the severity of the condition must be judged on objective findings During infancy the best



FIGURE XVII–17. Valvular pulmonary stenosis combined with subvalvular aortic stenosis (same patient as in Figure XVII–16). Child.

pulses were of good volume. The blood pressure was normal. The x ray showed great cardiac enlargement with a slightly upturned rounded apex; the pulmonary vascularity was normal or slightly reduced (see Figure XVII–17). The electrocardiogram showed a right axis deviation and evidence of right ventricular hypertrophy. It differed from the electrocardiogram usually seen in severe pulmonary stenosis in that V_1 showed deep S waves in addition to conspicuous R waves, and V_3 showed unusually high T waves. In this instance the electrocardiographic findings were the only ones which offered a clue to the existence of the aortic lesion (see Figure XVII–18).

TREATMENT

Medical treatment is indicated in an infant who develops cardiac failure in the neonatal period. The infant should rapidly receive his calculated dose of digitalis or one of the allied preparations. With such treatment he usually regains compensation and thereafter does well for many months and often for years. If he fails to respond to digitalis, immediate operation is indicated.

Furthermore, if the infant does well in the neonatal period and subsequently develops cardiac failure with a large pulsating liver, prompt operation is indicated. Indeed, the condition constitutes a veritable surgical emergency.

Digitalis, except in the neonatal period, is seldom indicated. If, however, the patient shows signs of cardiac failure, digitalis and diuretics are of value. Such treatment is purely palliative; prompt operation is imperative.

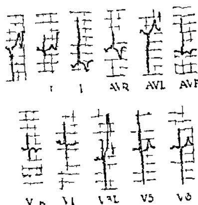


FIGURE XVII-18 Valvular pulmonary stenosis combined with subvalvular aortic stenosis (same patient as in Figure XVII-16) Child

Prophylactic antibiotics are indicated throughout life for the prevention of subacute bacterial endocarditis when there is danger of infection from dental extractions or other procedures Even after successful operation, the pulmonary valve is not normal the patient is still susceptible to the disease

Limitation of exercise is rarely necessary, as the condition can be relieved by surgery Therefore only if surgical treatment is unavailable is restriction of activity indicated In most parts of the world a child with a valvular pulmonary stenosis may be permitted to lead a normal life, if his activity places a strain on his heart, operation is indicated

The advisability of surgical treatment depends upon the severity of the stenosis In some instances the pulmonary stenosis is so slight that the heart is normal in size, the pulmonary blood flow can be increased with exercise, and the electrocardiogram does not even show evidence of right ventricular hypertrophy Such a patient is asymptomatic and, if he has attained his full growth, he will certainly remain so The condition need cause no concern The prognosis is excellent without operation

In the author s opinion if the heart remains normal in size and the patient is asymptomatic and able to lead a normal life and the pressure is under 100 mm of mercury in the right ventricle operation is not indicated, even if the electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy It must be remembered that operation does not restore the valve to normal and that mild pulmonary stenosis is compatible with a long and active life

Surgical treatment is indicated if there is evidence of severe or progressive cardiac strain Inasmuch as cardiac enlargement precedes symptoms, the severity of the condition must be judged on objective findings During infancy the best

guide is the size of the heart in the x ray If it is but slightly enlarged, serial x rays at monthly or bimonthly intervals will reveal whether or not the condition is progressive During childhood enlargement occurs more slowly, therefore the electrocardiogram offers the best index of the amount of work required of the right ventricle Regardless of the electrocardiogram, cyanosis which persists in childhood is always an indication for operation as it occurs only when the pressure in the right auricle is greater than that in the left auricle

In brief, *the indications for operation* are persistent cyanosis, or a right ventricular pressure of over 100 mm of mercury combined with evidence of right ventricular hypertrophy in the electrocardiogram or cardiac enlargement The ideal time for operation depends not on the age of the child, but on the size of the heart It is wise to operate prior to the occurrence of great cardiac enlargement or cardiac failure Therefore, if there is progressive increase in the size of the heart out of proportion to the growth of the patient, operation is indicated In young infants immediate operation is indicated if there is great cardiac enlargement or pulsations palpable at the margin of the liver

Several types of operation have been developed The first was that of valvulotomy developed by Sir Russel Brock,[1] who used the transventricular approach to cut the pulmonary valve and dilate the pulmonary orifice This operation has been simplified by Potts[12] by the invention of special instruments Swan and his associates[13] have advocated the use of hypothermia, so that the valve may be approached from the pulmonary artery and divided under direct vision Søndergaard[14] has developed a technique by which the operation may be accomplished through the pulmonary artery but without hypothermia Recently a number of surgeons have advocated operation under direct vision with the aid of a pump and oxygenator

At operation the surgeon is usually able to confirm the diagnosis by the finding of a large pulmonary artery in which the pressure is low Frequently, with each ventricular systole, he can feel the forceful thrust of the pulmonary valve against his finger, or he may feel the jet of blood forced through the stenosed pulmonary valve into the pulmonary artery, occasionally one can see a jet of blood as it impinges against the opposite wall of the pulmonary artery Furthermore, it is possible to measure the pressure in the right ventricle and in the pulmonary artery both before and after valvulotomy Measurement of the pressure after operation is of aid in order to make sure that the valve orifice is sufficiently enlarged so that the right ventricular pressure is reduced to normal or nearly to normal Nevertheless, if the pulmonary stenosis has been adequately relieved, failure of the right ventricular pressure to return to normal at operation is usually

not an indication for resection of the muscle at the base of the right ventricle In deed Engle et al[1] have shown that, after the relief of pulmonary stenosis, there may be a gradual drop in right ventricular pressure and over a period of months there may be a return to normal

In this operation the prime consideration is the relief of the pulmonary stenosis Even if prior to operation the patient has shown persistent cyanosis, when the pressure in the right auricle becomes normal, the valve covering the foramen ovale will clo e Therefore only if there is a gross defect in the auricular septum is closure of the defect necessary Unless the heart is greatly enlarged, the risk of operation is remarkably slight In 1953 the mortality rate, based on over 2,000 operations done in widely different places, was only 4 per cent

The results of operation can be judged by the change in the size of the heart in simple anterior posterior teleoroentgenograms and by the changes seen in the electrocardiogram Therefore cardiac catheterization is not necessary in order to determine the right ventricular pressure The more completely the pressure in the right ventricle is relieved, the more promptly the cardiac enlargement comes to an end[16] Except when the heart is greatly enlarged, if the right ventricular pres sure drops to normal, the heart returns to normal size Landtman[17] has shown that the electrocardiogram also offers evidence of the relief of right ventricular strain in that the axis becomes balanced and over a period of months right ven tricular dominance disappears Significant changes in the electrocardiogram are usually not apparent until six months after operation and it may be a year or longer before the maximum change is attained (see Figure xvii–19)

When the heart is enormously enlarged, the risk of operation is great and it is doubtful that perfection can be attained Nevertheless, the benefit may be tre mendous The patient usually regains and maintains compensation, dyspnea disappears and his exercise tolerance greatly increases With lesser degrees of enlargement the heart will return to normal and the patient will be able to lead an entirely normal life

It is however, important to remember that the heart itself is not entirely normal The fibrotic valve is only cut it is *not a normal valve* Resection of the infundibular area does not restore the heart to normal The patient will remain susceptible to subacute bacterial endocarditis, therefore throughout his life he should receive the usual prophylactic treatment

PROGNOSIS

The prognosis varies with the severity of the pulmonary stenosis Mild de grees of pulmonary stenosis are compatible with a long and active life

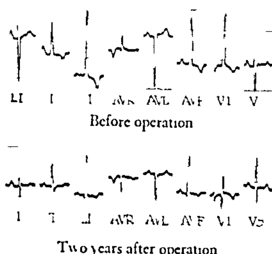


FIGURE XVII–19 Valvular pulmonary stenosis with an intact ventricular septum

Even moderate degrees of pulmonary stenosis may be compatible with longevity Currens et al[3] have reported the case of a man with severe pulmonary stenosis and patency of the foramen ovale who lived to be seventy years of age The author has seen a specimen of a pulmonary stenosis from a man who served in the regular Army, contracted rheumatic fever late in life, and died of rheumatic heart disease when nearly seventy years of age In this instance, in addition to mitral and aortic stenosis, autopsy revealed a cap like stenosis of the pulmonary valve which reduced the lumen to approximately one third of its normal diameter This degree of pulmonary stenosis had apparently never caused any difficulty

If the pulmonary stenosis is sufficiently severe to place a strain on the right side of the heart, the strain can be relieved by operation Since the development of cardiac surgery, most patients with this malformation may expect to enjoy long and active life

Severe pulmonary stenosis greatly increases the work of the right side of the heart and unless the condition is corrected by surgery it leads to progressive cardiac enlargement, engorgement and pulsation of the liver, and eventually to edema and ascites Patency of the foramen ovale lessens the strain on the heart but increases the oxygen unsaturation of the arterial blood and leads to polycythemia Even for individuals with severe pulmonary stenosis who have not sought medical attention until after the heart is markedly enlarged, the life span may be greatly lengthened by successful operation

SUMMARY

Pulmonary stenosis with an intact ventricular septum is a relatively common malformation Usually the stenosis is of the valvular type but in rare instances *there may* be an infundibular stenosis The pulmonary stenosis usually increases w[illegible] leads to progressive cardiac enlargement The obstruction to the pu[illegible]
the [illegible]
pressure in the right auricle rises if the valve covering the foramen ovale is not completely sealed, it will be forced open and a right to-left shunt will be established through it

The clinical findings vary with the severity of the pulmonary stenosis and with the structure of the foramen ovale

The patient is of a sturdy build

Dyspnea is the outstanding complaint Nevertheless, attacks of paroxysmal dyspnea are virtually unknown in infancy

The presence or absence of cyanosis depends upon the structure of the auricular septum If, as happens with 75 per cent of these patients, the foramen ovale is not completely sealed, cyanosis develops insidiously and generally becomes apparent between two and seven years of age

Polycythemia develops as the oxygen unsaturation of the arterial blood increases

Clubbing of the extremities gradually develops after cyanosis becomes apparent

The neck vessels may be engorged and show conspicuous pulsations

Enlargement of the liver occurs as the heart increases in size Pulsations at the margin of the liver are common, especially if the auricular septum is intact

Edema and ascites are late manifestations

Loss of consciousness is also a late manifestation

The size of the heart varies with the severity of the pulmonary stenosis With slight to moderate pulmonary stenosis there is no cardiac enlargement

Cardiac enlargement precedes symptoms therefore a patient suspected of this malformation should be kept under observation until he has attained his full growth Serial x rays give *the best evidence* of progressive cardiac enlargement in infancy whereas the electrocardiogram *is the most sensitive* index in childhood

A harsh systolic murmur and a thrill over the pulmonary area and in the

suprasternal notch, combined with a weak or absent second sound, are characteristic of this malformation

Infundibular stenosis is to be suspected if the murmur is characteristic but there is no thrill in the suprasternal notch, it is also to be suspected if the murmur is maximal in the third and fourth left interspaces, and the second sound at the base is clearly audible

The x ray shows fullness of the pulmonary conus and may show conspicuous pulsations in this area but minimal pulsations in the hilar shadows With tremendous cardiac enlargement in early infancy or in adults with infundibular stenosis, there may be no poststenotic dilatation and hence no fullness of the pulmonary conus

The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy which becomes pronounced as the strain on the right ventricle increases

The circulation time is usually prolonged

The oxygen saturation of the arterial blood varies with the amount of venous blood shunted through the foramen ovale to the systemic circulation

Cardiac catheterization reveals high pressure in the right ventricle and low pressure in the pulmonary artery

Angiocardiography shows prompt opacification of the pulmonary artery, in which the dye lingers for a long time The aorta is not visualized until after the left auricle and the left ventricle have been delineated

The diagnosis is based upon the finding of a harsh systolic murmur and a thrill, a weak or absent pulmonic second sound, and electrocardiographic evidence of a right axis deviation and right ventricular hypertrophy

The malformation calls for differentiation from an auricular septal defect, from primary pulmonary hypertension, occasionally from the Eisenmenger complex, and in some instances from a complete transposition of the great vessels, from a tetralogy of Fallot, and from defective development of the right ventricle combined with pulmonary stenosis

Medical treatment, especially digitalis, is of great benefit if an infant develops cardiac failure in the neonatal period Digitalis should also be given to any patient who develops cardiac failure before he can reach a hospital where cardiac surgery is performed Inasmuch as the condition can be relieved by surgery, limitation of exercise is seldom necessary

Mild degrees of pulmonary stenosis may be compatible with a long and active life, therefore in such instances operation is not indicated On the other hand,

severe degrees of pulmonary stenosis lead to progressive cardiac enlargement and cardiac failure with edema and ascites

Operation is indicated if there is persistent cyanosis, cardiac enlargement or evidence of right ventricular strain or excessively high pressure in the right ventricle

Several types of operation have been devised All the operations give excel ... [illegible] ...estored to normal, the risk of subacute [illegible] ... throughout his life

receive prophylactic chemotherapy throughout his life

For a patient who shows evidence of cardiac strain, operation removes the strain on the heart and changes the prognosis from bad to excellent

B *Stenosis in Either or Both Branches of the Pulmonary Artery*

In 1938 Oppenheimer[18] reported a case of stenosis of both main branches of the pulmonary artery and reviewed the literature No clinical attention was paid to the condition until 1954-55 In 1954 Søndergaard[19] reported the condition as coarctation of the pulmonary artery The following year Arvidsson et al [20] reported the occurrence of multiple stenoses of the pulmonary artery which they diagnosed by selective angiocardiography In the same year an additional case was reported by Kjellberg et al [21] In 1957 eight more clinical cases were reported by Gyllensward et al As in most instances, once an anomaly has been recognized it is detected with increasing frequency Furthermore, it produces a distinctive clinical syndrome

NATURE OF THE MALFORMATION

A localized constriction of the pulmonary artery may occur almost anywhere along its branches Gyllensward et al have emphasized that there are two main types Type I consists of a short membranous stenosis about 1 cm distal to the pulmonary valve and an elongated mild stenosis of one of the branches of the pulmonary artery and Type II is an extensive abnormality with multiple, short constrictions which usually involve the pulmonary arteries in both lungs Whether or not it is usual to have calcification in the pulmonary artery, as reported by Oppenheimer is not known, as most of the recent reports have been based on clinical observations and lack pathological confirmation

Actually there may be a single constriction in one main branch, bilateral constrictions in both main branches, or multiple constrictions scattered throughout

the branches of the pulmonary arteries Such constrictions affect the heart and circulation in a manner closely similar to that which occurs in valvular pulmonary stenosis, the only difference is that the obstruction in the pulmonary circulation lies beyond, instead of at, the pulmonary valve When the constriction is limited to one lung, it causes little or no strain on the circulation If, as more commonly occurs, it is bilateral, it places a strain on the circulation which is functionally similar to that caused by isolated valvular pulmonary stenosis Indeed, such a constriction of the pulmonary artery may occur in conjunction with valvular pulmonary stenosis Further, the abnormality may occur in combination with malformations which cause persistent cyanosis, under such circumstances each will contribute its own component It is the analysis of the separate components which will permit accurate diagnosis Hence this section is concerned with stenosis of the branches of the pulmonary artery when it occurs as an isolated abnormality

COURSE OF THE CIRCULATION

The course of the circulation is unaltered by the malformation unless the foramen ovale is held open by the high pressure in the right auricle Thus the course of the circulation is fundamentally the same as that of isolated valvular pulmonary stenosis (see Diagrams XVII-1 and 2)

PHYSIOLOGY OF THE MALFORMATION

The constriction of the pulmonary artery affects the circulation in a manner similar to that of any constriction elsewhere The pressure proximal to the obstruction is higher than that distal to it The narrowed area in the pulmonary artery obstructs the flow of blood to the lungs The pressure in the pulmonary artery proximal to the constriction is increased, hence more work is required of the right ventricle to pump the blood to the lungs Consequently there is right ventricular hypertrophy If the stenosis is sufficiently extreme, the high pressure in the right ventricle is transmitted back into the right auricle As the pressure in the right auricle rises, if the valve covering the foramen ovale is not completely sealed, it will be forced open and a right to-left shunt established at the auricular level

CLINICAL FINDINGS

The symptoms produced by this anomaly vary with the number and severity of the constrictions When there are bilateral constrictions in the pulmonary ar

teries, the clinical findings are similar to those in isolated valvular pulmonary stenosis

Cyanosis is usually absent If, however, the foramen ovale is not completely sealed when the pressure in the right auricle exceeds that in the left auricle, the valve will be forced open and a right to-left shunt established Such was the situation in the case reported by Oppenheimer

Dyspnea on exertion may occur because the obstruction to the pulmonary blood flow prevents the increase in the circulation to the lungs which normally occurs with exercise

CARDIAC FINDINGS

The heart may be normal in size and shape and there may be no murmurs over the precordium Since the work of the right ventricle is increased, the condition may lead to progressive cardiac enlargement. *The second sound* over the pulmonary area is loud and banging, similar to that which occurs in pulmonary hypertension

A continuous murmur is audible over the lungs It has the humming quality similar to that produced by an anastomosis between a systemic artery and one of the branches of the pulmonary artery The continuous murmur is due to the continuous flow of blood from the area of high pressure proximal to the constriction to the area of low pressure distal to it Hence it is better heard when there is but a single constriction in one or both of the main branches of the pulmonary artery than when there are multiple constrictions throughout the lungs Indeed under the latter circumstance, the continuous murmur may be absent because the pressure gradient is gradually reduced between each of the successive constrictions

X RAY AND FLUOROSCOPIC FINDINGS

The contour of the heart is similar to that of pure pulmonary stenosis The prominence of the pulmonary conus is, however, not caused by poststenotic dilatation but by dilatation of the pulmonary artery proximal to the stenosis The multiple constrictions in the branches of the pulmonary artery reduce the size of these vessels Hence the vascular markings are decreased and the lungs appear clear

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram shows evidence of the increased work required of the

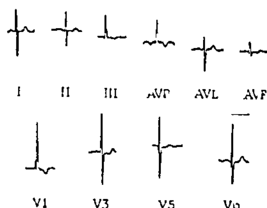


FIGURE XVII-20 Peripheral pulmonary stenosis (clinical diagnosis, not proven)

right ventricle The standard leads show a right axis deviation and the unipolar precordial leads show evidence of right ventricular hypertrophy (see Figure XVII-20)

SPECIAL TESTS

The red blood cell count, the amount of available hemoglobin, and the hematocrit reading are normal It is only in the rare instance when the foramen ovale is held open by the high pressure on the right side of the heart that the patient may develop polycythemia Usually, however, if polycythemia is present it is caused by some additional malformation

Cardiac catheterization shows an increased pressure in the right ventricle and in the pulmonary artery and no evidence of a shunt These findings are similar to those of primary pulmonary hypertension In order to demonstrate the constriction in the pulmonary artery the catheter must slip into the branch of the pulmonary artery in which the constriction lies and, furthermore, the constriction must not be so extreme that it is impossible to pass the catheter through it A number of investigators[20, 3, 4] have reported cases in which it was possible to demonstrate high pressure proximal to the constriction and an abrupt fall in the pressure when the catheter passed through the constriction

Angiocardiography may also reveal the condition[0] by the demonstration of abrupt narrowing in the branches of the pulmonary artery Proximal to the constriction the pulmonary artery and its branches are dilated and distended and there is an abrupt decrease in the size of the artery Arvidsson et al[0] have reported four cases diagnosed by selective angiocardiography

DIAGNOSIS

The diagnosis is based upon the clinical finding of a continuous murmur

over one or both lungs, combined with an accentuated pulmonic second sound It may be confirmed by cardiac catheterization or angiocardiography

DIFFERENTIAL DIAGNOSIS

The condition, when uncomplicated, may be confused with persistent patency of the ductus arteriosus, with primary pulmonary *hypertension*, with a hemi truncus arteriosus, or with other rare anomalies which cause a continuous murmur (see Chapter xxv)

Persistent patency of the ductus arteriosus causes a crescendo-decrescendo murmur of maximal intensity over the pulmonary area The second sound is audible within the continuous murmur In contrast to this, in the malformation under discussion, the pulmonic second sound is loud and banging and the continuous murmur is audible over the lungs

Primary pulmonary hypertension may be confused with this malformation because of the absence of murmurs over the precordium and the marked accentuation of the second sound over the pulmonary area, combined with evidence of high pressure in the right ventricle and in the pulmonary artery The presence of a continuous murmur over the lungs differentiates the two conditions

A hemi truncus arteriosus may readily be confused with localized constrictions in the pulmonary artery Both are rare anomalies which cause a continuous murmur over the lungs A hemi truncus arteriosus causes no strain on the right side of the heart (see Chapter xiv, Section B) Indeed, the right ventricle pumps the blood to only one lung whereas when the pulmonary artery is constricted, the work of the right ventricle is increased Hence the electrocardiogram aids in the differentiation of the two conditions

The malformation under discussion must also be differentiated from the other rare anomalies which cause a continuous murmur (see under Differential Diagnosis in Chapter xxv)

When there is persistent cyanosis this malformation calls for differentiation from a truncus arteriosus with relatively adequate pulmonary blood flow Cardiac catheterization or angiocardiography shows that the pulmonary artery, not the aorta or the truncus, arises from the right ventricle and there is no evidence of a shunt in the right ventricle

TREATMENT

As of 1960 there has been no *satisfactory treatment* Although in *theory* the constricted area could be resected as it is in coarctation of the aorta, the occur

rence of multiple constrictions renders such an operation difficult if not impossible

PROGNOSIS

The prognosis depends upon the severity of the condition Although the author has seen one baby with severe bilateral constrictions in the main branches of the pulmonary artery who died of right sided cardiac failure in early infancy,[18] the condition is generally far less severe and is usually compatible with relative longevity

SUMMARY

Stenosis may occur in either or both branches of the pulmonary artery There may be single or multiple stenoses Severe constriction of the pulmonary artery increases the pressure against which the right ventricle must work The condition may lead to progressive right sided cardiac enlargement The second heart sound over the pulmonary area is accentuated and there may be a continuous murmur audible over the lungs The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy Cardiac catheterization shows high pressure in the right ventricle and in the main pulmonary artery If it is possible to pass the catheter through the constriction, the pressure distal to the constriction will be lower than that proximal to it It may be possible to demonstrate the area of constriction by angiocardiography

The condition requires differentiation from persistent patency of the ductus arteriosus, pulmonary hypertension, a hemi truncus arteriosus, and other causes of a continuous murmur Multiple peripheral pulmonary stenoses are difficult to correct

The prognosis varies with the severity of the condition When the stenosis is severe, the prognosis is guarded Less severe stenoses are compatible with relative longevity

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CHAPTER XVIII

PULMONARY HYPERTENSION

PULMONARY hypertension may be primary or it may be 'secondary' Primary pulmonary hypertension means that the initial pathology lies in the pulmonary vascular bed and that the heart is normal except for the strain placed on it by the increased peripheral resistance in the *pulmonary* vascular bed In contrast to this, secondary pulmonary hypertension *means* that the high pressure in the pulmonary artery is *secondary to a malformation* of the heart Edwards[1] has pointed out that in a number *of malformations* of the heart the blood is ejected to the lungs under systemic pressure Under such circumstances the *high* pressure in the *lungs is secondary* to the cardiac malformation A brief review of the normal changes in the lungs which occur immediately after birth will help to clarify the nature of this pathology

Civin and Edwards made a detailed study of microscopic sections of the lungs of patients from birth to old age These authors followed Brenner's classifications of the division of the intrapulmonary arteries into three main groups (1) elastic arteries, (2) muscular arteries, and (3) arterioles The investigators demonstrated that in the fetus the lumen of muscular arteries of the lungs is narrow and the media is thick, the adventitia is thick and the intima thin Normally during the first six months of extra uterine life the lumen of the muscular arteries widens and the media becomes thin Thus the lumen of the arteries increases in size and the thickness of the muscular wall decreases The adventitia becomes much thinner and the intima remains unchanged Such are the normal changes which occur during the first months of life Moreover, the pulmonary *vascular bed continues* to expand up to about the twentieth year of life The *greatest changes, however, normally* occur in early infancy

Edwards has postulated that, since no well-established *vasomotor response* in the pulmonary vascular bed has been demonstrated, the *narrow* lumina of the muscular arteries constitute one factor which increases the *resistance* to the pulmonary flow during fetal life, and, furthermore, that the gradual increase in the size of these lumina lowers the pulmonary resistance He has found *that the* principal changes in the pulmonary vascular bed which occur in the *first* few months of extra uterine life are the widening of the *lumina* of the muscular arteries and the thinning of the media of these vessels

Furthermore, Edwards has pointed out that in a number of malformations, mainly those in which the blood is pumped into the pulmonary artery under systemic pressure, the pressure in the pulmonary artery is a vital factor in the distribution of the blood to the two circulations Were the pulmonary pressure low, the pulmonary blood flow would be so excessive that the patient would drown in his lungs Edwards has reported narrowing of the intrapulmonary vessels in a patient with an Eisenmenger complex[1, 3] and also in two patients in whom the ductus arteriosus opened into the descending aorta distal to the coarctation[4] The muscular arteries were characterized by a thick media and a relatively narrow lumen In older patients he has observed that, in addition to the thick media, intimal fibrosis develops He believes that, in malformations of the heart in which there is excessive pulmonary blood flow, the muscular arteries maintain their fetal characteristics of a small lumen and a thick media as a compensatory mechanism These changes initially are caused by a delay in the opening up of the pulmonary vascular bed which protects the lungs from an excessive pulmonary blood flow under abnormally high pressure Edwards conceives that the initial changes are reversible in that, if the excessive pressure were removed, the lungs would be capable of normal expansion The long-continued high pressure ultimately leads to intimal proliferation and fibrosis These changes rigidly fix and further constrict the lumina of the vessels and thus further increase the resistance to the pulmonary blood flow When such changes occur, the condition is no longer reversible Thus, although in the early stages pulmonary hypertension is compensatory and the changes are reversible, in the later stages changes occur which are progressive and irreversible

Edwards has presented strong evidence to show that such is the sequence of events in malformations of the heart in which blood is ejected simultaneously into the systemic circulation and the pulmonary circulation Pulmonary hypertension always occurs in an Eisenmenger complex, in a single ventricle without pulmonary stenosis, in a truncus arteriosus in which large pulmonary arteries arise from the base of the truncus, and in a Taussig Bing heart Whenever the right ventricle pumps the blood to the systemic circulation through the ductus arteriosus, the pressure in the pulmonary artery must be the same as that in the systemic circulation, such is the situation in aortic atresia or marked hypoplasia of the ascending aorta when the systemic circulation is supplied from the pulmonary artery through the ductus arteriosus

In addition to these conditions in which pulmonary hypertension is in a sense an integral part of the malformation of the heart, or at least is the inevitable re

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tion quite as profoundly as does a coarctation of the ... ence is that the pulmonary hypertension places a strain on the right side of the heart, whereas coarctation of the aorta places a strain on the left side

ETIOLOGY

The etiology is obscure Indeed, the author believes that there may be a number of different causes of "primary" pulmonary hypertension In some instances the increased pulmonary pressure appears to be secondary to some pulmonary infection or to the inhalation of some irritating substance In other instances multiple thrombi have been found in the smaller pulmonary vessels and occasionally there is a massive thrombus in the main branches of the pulmonary artery In still other instances it appears to be secondary to long standing pulmonary disease and asthma Pulmonary hypertension is also seen in severe kyphoscoliosis It is possible that there may be a pulmonary arteriolar disease which is comparable to systemic arteriolar sclerosis Cor pulmonale may appear as an acute disease or as a chronic illness The disease is notoriously common in and around Pittsburgh, where it seems as if it were an occupational disease It is also known to occur in families, as for example in the family referred to the author by Dr Dresdale,[5] where the mother, her sister, and her son all died of pulmonary hypertension In the family reported by Coleman et al [6] three siblings suffered from "primary" pulmonary hypertension Furthermore, the author has studied patients of all ages, from six months to over twenty years, with what appeared to be "primary" pulmonary hypertension

The pathology in the lungs is not the same in all instances There is one group of cases in which the minute vessels in the capillaries of the lungs show an endarteritis and inflammatory changes In another group of cases the lesions appear to lie in the small muscular arteries and numerous thrombi occur in the lungs, many of which show evidence of recanalization These changes are similar to those described by Rich[7] in patients with long standing polycythemia but have also been observed in a number of infants in whom there was no polycythemia Such lesions are being reported with increasing frequency in adults with cor pulmonale

It is also conceivable that 'primary' pulmonary hypertension may be due to an arrest in the development of the pulmonary vascular bed of such a nature that the intrapulmonary arteries fail to undergo their normal postnatal involution It may be that, in the presence of a congenital abnormality of the pulmonary vascular bed, there are two opposing factors one is an abnormality of the lungs which is of such a nature as to increase the peripheral resistance and the other is the gradual expansion of the pulmonary vascular bed which, as Edwards has shown, normally continues up to twenty years of age Thus, during childhood and adolescence, the latter may offset the former with the result that the increased pulmonary resistance may not become manifest until early adult life Furthermore, it is possible that in some instances an acquired pulmonary infection which would produce insignificant scarring in a normal adult may be sufficient to swing the balance unfavorably in a patient with an abnormality in the pulmonary vascular bed

It is notoriously true, and has been repeatedly emphasized, that although patients with cor pulmonale give a history of repeated respiratory infections, the condition is rarely seen in chest clinics among the patients who are known to suffer from long standing or chronic pulmonary infections

Allergy appears to be a factor in some instances, as in one of the author's patients who was extremely short of breath, so short of breath that she panted over the telephone After a few weeks in Maine this patient was able to enjoy swimming and, when she returned, the dyspnea was no longer detectable over the telephone

Although it has long been assumed that there is no nervous control of the lungs, there is increasing evidence that such a mechanism may exist Lilienthal and Riley,[8] in their review of diseases of the respiratory system, referred to the possibility and discussed the evidence in favor of it Indeed, the response of older patients with an Eisenmenger complex to the inhalation of oxygen also strongly suggests the existence of some nervous or humoral control over the pulmonary vascular bed

As previously mentioned, the author has studied a number of infants who suffered from primary" pulmonary hypertension When the difficulty dates from birth, it seems clear that the pulmonary hypertension is congenital in origin Thus it appears that pulmonary hypertension may be congenital or may result from acquired disease

Regardless of etiology, 'primary' pulmonary hypertension occurs as an iso-

lated abnormality and may be present from birth The increased resistance in the pulmonary vascular bed increases the work required of the right ventricle and this in turn increases the work required of the right auricle As in all conditions which place a strain on the right side of the heart, the foramen ovale may be held open by the high pressure in the right auricle In contrast to the abnormality in valvular pulmonary stenosis with an intact ventricular septum, and in Ebstein s anomaly of the tricuspid valve, the difficulty lies not within the heart but in the lungs, consequently the high pressure in the pulmonary artery may tend to keep the ductus arteriosus from closing Thus 'primary pulmonary hypertension occurs as an isolated anomaly and it also occurs with persistent patency of the ductus arteriosus, the latter combination of anomalies produces a distinctive clinical syndrome and is discussed in Section B Section A is concerned with primary pulmonary hypertension with and without patency of the foramen ovale

A *Primary Pulmonary Hypertension with and without Patency of the Foramen Ovale*

NATURE OF THE MALFORMATION

This malformation is primarily due to changes in the pulmonary vascular bed The peripheral pulmonary vascular bed is abnormally constricted and thereby the peripheral resistance in the lungs is increased and the intrapulmonary arterial pressure is abnormally high The heart itself is normally formed The ductus arteriosus is closed Although the nature of the changes in the lungs is not fully understood, and indeed may not always be the same, any condition which increases the peripheral resistance in the pulmonary vascular bed increases the work of the right ventricle The right ventricle becomes slightly dilated and greatly hypertrophied The strain on the right ventricle may be so great that the pulmonary valve becomes insufficient. The increased pressure in the right ventricle increases the work required of the right auricle to pump the blood into the right ventricle consequently the right auricle is also hypertrophied Inasmuch as the heart is normal, if the foramen ovale has not been completely sealed, the valve will be forced open by the high pressure in the right auricle and thereby a right to-left shunt will be established This occurs in approximately 75 per cent of all patients in whom the pressure in the right auricle is abnormally high during early infancy

COURSE OF THE CIRCULATION

If the foramen ovale becomes completely sealed and the ductus arteriosus undergoes normal obliteration, the course of the circulation is normal The blood flows from the right auricle to the right ventricle and thence is pumped out through the pulmonary artery to the lungs The constriction of the pulmonary vascular bed increases the peripheral resistance in the lungs and this in turn increases the work of the right side of the heart Nevertheless, the blood which passes through the lungs is fully oxygenated and is returned in the normal fashion to the left auricle, thence it flows into the left ventricle and is pumped out through the aorta to the systemic circulation It follows that the blood in the systemic circulation is fully oxygenated The blood is returned in the normal manner by the superior vena cava and the inferior vena cava to the right auricle, there the cycle starts again (see Diagram XVIII–1)

As with all malformations which place a strain on the right side of the heart, as the pressure in the right ventricle rises, the right auricle cannot empty itself as readily as normally Consequently, if the foramen ovale is not completely sealed, as the pressure within the right auricle rises, the increased pressure in that chamber eventually will force the valve covering the foramen ovale to open and some venous blood from the right auricle is shunted into the left auricle Under such circumstances the mixture of oxygenated blood which is returned to the left auricle from the lungs and of venous blood which is shunted from the right auricle to the left auricle, will flow into the left ventricle and be pumped out through the aorta into the systemic circulation The oxygen saturation of the arterial blood will be reduced directly in proportion to the volume of venous blood which is shunted from the right auricle through the foramen ovale to the systemic circulation When the volume of unoxygenated blood so shunted becomes sufficiently great, the patient will show cyanosis (see Diagram XVIII–2)

PHYSIOLOGY OF THE MALFORMATION

The physiology of the malformation concerns the abnormality in the lungs There is progressive narrowing of the small arteries and arterioles in the pulmonary vascular bed The pulmonary capillary bed usually remains normal and hence the pressure in the capillaries remains normal Nevertheless, the increased resistance in the pulmonary vascular bed increases the work required of the right ventricle and consequently increases the pressure in that chamber The increased pressure in the right ventricle increases the pressure against which the right auricle must work and this in turn raises the pressure in that chamber If the fora

men ovale is not completely sealed, it acts as an escape valve and slightly reduces the strain on the heart *If it is entirely* sealed, the escape valve is closed and cardiac failure occurs early The peripheral resistance is frequently so greatly increased that the pressure in the pulmonary circulation exceeds the systemic pressure Unless the pressure can be relieved the condition leads inevitably to progressive right sided cardiac enlargement and cardiac failure

CLINICAL FINDINGS

The clinical findings vary with the severity of the condition In some instances the condition may be so severe that the infant dies of cardiac failure before one year of age, in others the condition may be compatible with life for more than thirty years

Difficulty in feeding and *failure to gain* are common complaints in an infant with severe pulmonary hypertension

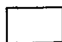
Repeated respiratory infections bronchitis and *pneumonia* frequently occur during infancy and childhood The patient may, however, be asymptomatic in infancy and childhood and no abnormality may be suspected until early adult life, when he suffers from some pulmonary infection and thereafter develops *dyspnea*

Dyspnea is the outstanding complaint In infancy rapid respiration is the initial manifestation Not infrequently the condition progresses to cardiac failure before the seriousness of the respiratory distress is appreciated In an older patient the onset of dyspnea may occur abruptly after an intercurrent pulmonary infection and become rapidly worse until he is so short of breath that conversation causes him to pant

Attacks of suffocation occasionally occur The pulmonary artery may be so large and the pressure in it so high that either of its main branches may compress the bronchi The author has seen one child in whom the compression was so severe that he suffered from severe dyspnea after lying for a short time flat in bed Bronchoscopic examination revealed compression of the right or left main bronchus, depending upon which side the child was lying

The presence or absence of cyanosis depends upon the structure of the foramen ovale Infants with severe pulmonary hypertension almost always show persistent cyanosis and oxygen unsaturation of the arterial blood, because the high pressure in the right auricle tends to keep the foramen ovale open and a right to left shunt is thereby established If the foramen ovale is completely sealed, the patient will show no cyanosis The oxygen saturation of the arterial blood is

DIAGRAM XVIII–1


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

DIAGRAM XVIII-1

Primary pulmonary hypertension

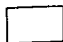
In this malformation the primary abnormality is the increased resistance in the pulmonary vascular bed the ductus arteriosus is obliterated and the foramen ovale is sealed The circulation of the blood is normal

The blood from the right auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs, where it is oxygenated The blood is returned in the normal manner to the left auricle thence it flows to the left ventricle and is pumped out to the systemic circulation and returned by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again

The only cardiac change which occurs is the hypertrophy of the right ventricle and the right auricle which is secondary to the increased work required of the right side of the heart to pump the blood to the lungs against the increased resistance in the pulmonary vascular bed

Clinical diagnosis Dyspnea is the outstanding complaint The heart is usually not greatly enlarged The pulmonic second sound is markedly accentuated There may be a loud early diastolic murmur The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy

Diagram XVIII–2


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis

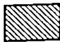

Venous blood

Diagram XVIII-2

Primary pulmonary hypertension with patency of the foramen ovale

In this malformation primary pulmonary hypertension is combined with patency of the foramen ovale. The foramen ovale acts as an escape valve for the high pressure in the right auricle by the establishment of a right to left shunt

Under such circumstances although most of the blood in the right auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs and is returned in the normal fashion to the left auricle some blood from the right auricle is shunted through the foramen ovale to the left auricle Consequently the left auricle receives fully oxygenated blood from the lungs and some venous blood from the right auricle This mixture of oxygenated and venous blood flows into the left ventricle and is pumped out through the aorta to the systemic circulation and is returned by the superior and inferior venae cavae to the right auricle There the cycle starts again

Clinical diagnosis Cyanosis usually develops insidiously It depends on the volume of venous blood shunted into the systemic circulation, hence cyanosis may be apparent at birth or may not develop until adolescence or adult life The heart is of approximately normal size with an insignificant systolic murmur and a markedly accentuated second sound over the pulmonary area The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy

approximately normal In an older patient who shows arterial oxygen unsaturation it may seem plausible that the unsaturation could be caused by the pathology in the lungs, actually in almost every instance in which a patient has shown cyanosis, autopsy has revealed patency of the *foramen ovale* Therefore it seems probable that the cyanosis in primary pulmonary hypertension has the same origin as that in "pure" pulmonary stenosis and in Ebstein's anomaly of the tricuspid valve namely, the cyanosis is caused by the shunting of venous blood from the right auricle through the *foramen ovale* to the systemic circulation

Clubbing is less marked than is the cyanosis

Polycythemia develops gradually as the patient suffers from persistent oxygen unsaturation of the arterial blood

Hemoptysis is a common complaint in older patients but seldom if ever occurs in infancy

The blood pressure is normal The *pulse* is of equal strength in the arm and the leg

Engorgement of the liver occurs as the heart begins to fail The liver may become enormously enlarged and frequently pulsations are palpable at its margin Engorgement of the liver occurs readily in patients who show no cyanosis, because the foramen ovale is completely or nearly completely sealed

Edema of the extremities is also a late manifestation Inasmuch as the condition is progressive, the edema may be extremely troublesome and difficult to treat

Ascites is also a late manifestation and may become extreme

Syncope is a late and serious manifestation The circulation through the lungs is so slow that the minute output of the heart is reduced Consequently there is not only a low pulmonary blood flow but also a low systemic flow Finally, the circulation of the blood becomes so sluggish that the supply of oxygen to the brain is seriously impaired Such appears to be the cause of the attacks of syncope from which these patients suffer After the development of syncopal attacks the duration of life is relatively short Many a patient dies during an attack of syncope

Precordial pain may occur but is rare It, too, is a late manifestation and is generally considered to be due to coronary insufficiency, it goes hand in hand with myocardial failure

CARDIAC FINDINGS

The size of the heart varies with the severity of the condition and the age at which it develops When severe pulmonary hypertension dates from birth, the

heart undergoes progressive enlargement and cardiac failure occurs at an early age When the pulmonary hypertension is less severe or develops at a later age, the heart may be but slightly enlarged Over a period of years the heart undergoes gradual progressive enlargement The pulmonary vascular disease may develop so rapidly that the condition is fatal before the heart becomes enormously enlarged

The second sound at the base of the heart to the left of the sternum is markedly accentuated and often reduplicated A decrease in the intensity of the second sound may, however, occur in the presence of cardiac failure, it is indicative of myocardial weakness

A systolic murmur may or may not be present Usually it is not loud

Pulmonary insufficiency frequently develops in children and young adults Indeed, the finding of a loud early diastolic murmur along the left sternal border in a patient with a markedly accentuated pulmonic second sound should always arouse suspicion of pulmonary hypertension The intensity of the diastolic murmur varies greatly with the patient s state of compensation If the condition is not too severe, a brief period of bed rest may cause the murmur to disappear, as the disease progresses, the patient may develop a systolic as well as a diastolic murmur The author has studied one seven year-old child in whom the murmur closely resembled the harsh continuous murmur of a patent ductus but was maximal in the third left interspace After two days of digitalis and rest in bed the continuous murmur disappeared and was replaced by an inconstant diastolic murmur There are few, if any, other cardiac abnormalities which show such variability of murmurs over such a short period of time

X RAY AND FLUOROSCOPIC FINDINGS

The heart may or may not be enlarged The pulmonary conus is full and usually the main pulmonary arteries are markedly enlarged Figure XVIII-1 shows great cardiac enlargement in a child of nine years Figure XVIII-2 shows the contour of the heart of a boy of thirteen with severe primary' pulmonary hypertension Fluoroscopy usually reveals a slight hilar dance and the peripheral lung fields are clear Examination in the oblique positions gives little additional information There is no enlargement of the left auricle, hence the esophagram is normal

ELECTROCARDIOGRAPHIC FINDINGS

The standard leads show a marked right axis deviation and the unipolar pre



FIGURE XVIII-1 Primary pulmonary hypertension Child

cordial leads show evidence of right ventricular hypertrophy or the pattern of so-called right ventricular "strain" (see Figure XVIII-3)

SPECIAL TESTS

The circulation time is normal or prolonged

Cardiac catheterization reveals high pressure on the right side of the heart The pressure in the right ventricle is elevated and the pressure in the pulmonary artery is as high or higher than that in the right ventricle but the pulmonary capillary pressure is usually normal If the foramen ovale is closed, there is no shunt Under such circumstances the oxygen content of the blood samples taken from the right auricle, the right ventricle, and the pulmonary artery should be identical Even if the foramen ovale is patent, the shunt is from right to left, there should be no significant difference in the oxygen content of the blood in the various places in the right side of the heart Cardiac catheterization is, however, not without risk, as these patients are extremely susceptible to arrhythmias



FIGURE XVIII–2 Primary pulmonary hypertension Child

Nevertheless, the procedure is more informative and less dangerous than angiocardiography

Angiocardiography is not helpful and may be dangerous The constriction in the pulmonary arteriolar bed slows the circulation of the blood through the lungs If the pulmonary hypertension is marked, the blockage of the pulmonary arterioles with the dye may entirely cut off the supply of oxygen to the patient and lead to death The only positive finding upon angiocardiography is the lingering of the contrast substance in the right ventricle and the main pulmonary artery Thus the picture is identical with that of valvular pulmonary stenosis with an intact ventricular septum

DIAGNOSIS

The diagnosis is based on the marked accentuation of the second sound at the base of the heart to the left of the sternum combined with evidence of right ventricular hypertrophy and strain The occurrence of a loud early diastolic murmur is common The x ray shows fullness of the pulmonary conus and the

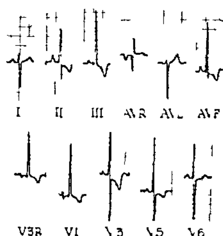


FIGURE XVIII-5 Primary pulmonary hypertension Child

main pulmonary arteries, upon fluoroscopy a hilar dance is visible Cardiac catheterization reveals a high pressure in the main pulmonary artery and no evidence of an intracardiac shunt or abnormality

DIFFERENTIAL DIAGNOSIS

In young infants who show cyanosis, the condition may simulate a tetralogy of Fallot, a complete transposition of the great vessels, or "pure" pulmonary stenosis, that is, valvular pulmonary stenosis in which the ventricular septum is intact and the foramen ovale is held open by the high pressure on the right side of the heart

In children and young adults, prior to the development of cyanosis, the condition may be confused with idiopathic dilatation of the pulmonary artery, a corrected transposition of the great vessels, an auricular septal defect, a patent ductus arteriosus without a continuous murmur, or acquired heart disease with *rheumatic or syphilitic aortic insufficiency, and occasionally with aortic stenosis* When the onset of cyanosis is delayed, the condition requires differentiation from polycythemia vera and the Eisenmenger complex With or without cyanosis the malformation must be differentiated from 'pure' pulmonary stenosis or Ebstein s anomaly of the tricuspid valve The condition may occasionally be confused with mitral stenosis and with a tumor of the left auricle

Tetralogy of Fallot may be confused with primary pulmonary hypertension in young infants who show deep persistent cyanosis and no murmurs Over a period of months, the evidence of progressive cardiac enlargement clearly differentiates the *malformation under discussion from a tetralogy of* Fallot

A complete transposition of the great vessels may also be confused with pri

mary pulmonary hypertension, as at birth in both instances the heart is normal in size and the vascular markings are slightly increased If there is pulmonary hypertension, as the heart undergoes enlargement the vascular markings decrease and the fullness of the pulmonary conus becomes apparent, thereby differentiating the condition from a complete transposition of the great vessels

Pure pulmonary stenosis resembles primary pulmonary hypertension in that progressive right sided cardiac enlargement and fullness of the pulmonary conus may occur, and there may or may not be cyanosis In pure pulmonary stenosis the systolic murmur is loud and harsh and ends abruptly with the end of systole and the second sound is weak or absent, whereas in primary pulmonary hypertension the second sound over the pulmonary area is markedly accentuated Indeed, it is only in the presence of cardiac failure and the consequent weakening of the second sound that the two conditions may be confused Under such circumstances cardiac catheterization may be necessary to establish the diagnosis

Idiopathic dilatation of the pulmonary artery differs from primary pulmonary hypertension in that the pulmonic second sound is not accentuated The electrocardiogram shows no evidence of extreme right ventricular hypertrophy Cardiac catheterization confirms the clinical impression that the pulmonary pressure is normal

Corrected transposition of the great vessels may be confused with 'primary' pulmonary hypertension because of the finding of marked accentuation of the second heart sound over the pulmonary area combined with x ray evidence of fullness of the pulmonary conus In a corrected transposition of the great vessels the electrocardiogram shows a tendency to a left axis deviation and evidence of left ventricular hypertrophy Furthermore, angiocardiography clearly shows that the fullness of the pulmonary conus is caused by the ascending aorta

An auricular septal defect is usually associated with a systolic murmur over the precordium which is frequently well heard posteriorly The pulmonic second sound is reduplicated but it is not as markedly accentuated as in patients with primary pulmonary hypertension The electrocardiogram shows a tendency to a right axis deviation, and commonly shows evidence of a right bundle branch block, rather than the pattern of extreme right ventricular hypertrophy

A patent ductus arteriosus may be mistaken for primary pulmonary hypertension especially in young infants while the pulmonary pressure remains high, that is before the development of a continuous murmur The heart is usually markedly enlarged, a systolic murmur and often a low pitched mid diastolic

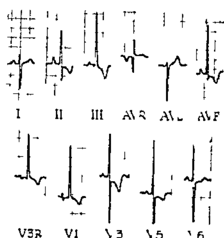


FIGURE XVIII–3 Primary pulmonary hypertension Child

main pulmonary arteries, upon fluoroscopy a hilar dance is visible Cardiac catheterization reveals a high pressure in the main pulmonary artery and no evidence of an intracardiac shunt or abnormality

DIFFERENTIAL DIAGNOSIS

In young infants who show cyanosis, the condition may simulate a tetralogy of Fallot, a complete transposition of the great vessels, or "pure' pulmonary stenosis, that is, valvular pulmonary stenosis in which the ventricular septum is intact and the foramen ovale is held open by the high pressure on the right side of the heart

In children and young adults, prior to the development of cyanosis, the condition may be confused with idiopathic dilatation of the pulmonary artery, a corrected transposition of the great vessels, an auricular septal defect, a patent ductus arteriosus without a continuous murmur, or acquired heart disease with rheumatic or syphilitic aortic insufficiency, and occasionally with aortic stenosis When the onset of cyanosis is delayed, the condition requires differentiation from polycythemia vera and the Eisenmenger complex With or without cyanosis the malformation must be differentiated from 'pure pulmonary stenosis or Ebstein's anomaly of the tricuspid valve The condition may occasionally be confused with mitral stenosis and with a tumor of the left auricle

Tetralogy of Fallot may be confused with primary pulmonary hypertension in young infants who show deep persistent cyanosis and no murmurs Over a period of months, the evidence of progressive cardiac enlargement clearly differentiates the malformation under discussion from a tetralogy of Fallot

A complete transposition of the great vessels may also be confused with pri

sounds are weaker and more confused The electrocardiogram does not show a marked right axis deviation and seldom shows evidence of right ventricular hypertrophy in V_1

Mitral stenosis is occasionally so severe that too little blood is forced through the mitral valve to cause a presystolic murmur Only a faint systolic murmur is audible over the base of the heart and the pulmonary second sound is accentuated X ray or fluoroscopic examination should reveal enlargement of the left auricle The electrocardiogram usually shows notched P waves associated with left auricular enlargement, and not the "pyramidal" P waves seen with dilatation and hypertrophy of the right auricle Cardiac catheterization not only shows high pressure in the right ventricle and in the pulmonary artery but also reveals a high "wedge" pressure

A tumor of the left auricle is rare Nevertheless, since the clinical syndrome[8] produced thereby resembles mitral stenosis, it too may require differentiation from primary pulmonary hypertension A tumor of the left auricle which obstructs the flow of blood through the mitral valve may give the clinical and laboratory signs of mitral obstruction without circulatory signs of mitral stenosis Fatigue is a common complaint There may be dyspnea on exertion and acute attacks of pulmonary edema Furthermore, the obstruction to the flow of blood through the left auricle causes back pressure in the lesser circulation, this eventually increases the pressure in the pulmonary artery and causes right sided cardiac strain Cardiac catheterization shows not only high pressure in the pulmonary artery but increased pressure in the pulmonary capillary bed Angiocardiography may be of aid in that it may demonstrate a filling defect in the left auricle

COMPLICATIONS

The complications result from the low pulmonary blood flow with the correspondingly low systemic flow and from the polycythemia As the circulation to the lungs becomes extremely sluggish, thrombosis may develop in the main branches of the pulmonary artery Such thrombi may become so enormous that they seriously obstruct the pulmonary blood flow and thereby still further increase the work required of the right ventricle Thrombosis of one of the main branches of the pulmonary artery should be suspected when there is enormous dilatation of the hilar vessels and minimal or no pulsation visible therein

Terminally the systemic circulation may become so sluggish that multiple systemic thrombi develop

murmur are present The lungs appear extremely vascular There is usually either a balanced electrocardiogram or a tendency to a left axis deviation with evidence of "combined" ventricular hypertrophy

Rheumatic or syphilitic heart disease and aortic insufficiency are considered only because of the occurrence of a diastolic murmur along the left sternal border The absence of any history of acute rheumatic fever or of syphilitic infection, combined with electrocardiographic evidence of right rather than left ventricular hypertrophy, should offer the clue to the diagnosis Furthermore, fluoroscopic examination will reveal *no fullness of the pulmonary conus* and slight, if any, hilar pulsations In doubtful cases, cardiac catheterization may be of help In the presence of aortic insufficiency the pressure in the right ventricle and in the pulmonary artery will be normal or moderately elevated, secondarily to back pressure from the left side of the heart, whereas with 'primary pulmonary hypertension the pressure in the right ventricle and in the pulmonary artery will be markedly elevated

Aortic stenosis may be considered because of the attacks of syncope and precordial pain The two conditions are, however, readily differentiated, as aortic stenosis places a severe strain on the left ventricle, whereas in 'primary" pulmonary hypertension the strain falls on the right ventricle In the former the electrocardiogram shows evidence of extreme left ventricular hypertrophy, whereas in the latter there is extreme right ventricular hypertrophy

Polycythemia vera does not cause oxygen unsaturation of the arterial blood The electrocardiogram is usually normal, whereas electrocardiographic evidence of a right axis deviation and right ventricular hypertrophy is the rule in the malformation under discussion Furthermore, in polycythemia vera cardiac catheterization reveals a normal pressure in the pulmonary artery

√*An Eisenmenger complex* differs from primary pulmonary hypertension in that a systolic murmur is more common and the second sound to the left of the sternum is not as forceful as in primary pulmonary hypertension The electrocardiogram shows greater evidence of left ventricular hypertrophy Fluoroscopy often reveals evidence of left auricular enlargement The circulation time is short There is oxygen unsaturation *of the arterial blood, which increases with* exercise Cardiac catheterization generally shows evidence of both a left to-right and a right to left shunt at the ventricular level

Ebstein's anomaly of the tricuspid valve can usually be differentiated from "primary" pulmonary hypertension in that the heart is larger and the heart

bed The increased resistance in the pulmonary vascular bed increases the work of the right side of the heart, there results right ventricular hypertrophy and dilatation of the main pulmonary artery and its branches

The course of the circulation is normal, except when the foramen ovale is held open by the high pressure in the right auricle and a right to-left shunt is thereby established

Dyspnea is the outstanding complaint

Engorgement of the liver and ascites develop as cardiac failure becomes severe

Syncope and precordial pain are late manifestations

The heart undergoes progressive enlargement, which is mainly right sided The pulmonic second sound is accentuated and there may be a long, loud, early diastolic murmur

Fluoroscopy reveals a fullness of the pulmonary conus, there may be a conspicuous hilar dance but the periphery of the lungs is clear

The electrocardiogram shows marked right axis deviation and evidence of right ventricular hypertrophy

The diagnosis is based upon the finding of a right sided cardiac enlargement and a markedly accentuated second sound over the pulmonary area The size of the heart, the age at which the patient develops symptoms, and the rapidity with which these advance to cardiac failure depend upon the etiology and the severity of the pulmonary vascular changes

The diagnosis is definitely established by cardiac catheterization and the finding of a high pressure in the pulmonary artery, with a normal wedge pressure, and no evidence of intracardiac shunt

The malformation must be differentiated from other conditions which cause right sided cardiac strain with or without persistent cyanosis In early infancy the condition may be confused with a tetralogy of Fallot or a complete transposition of the great vessels As the heart enlarges, its contour becomes similar to that of a pure pulmonary stenosis In the absence of cyanosis, the condition is most commonly confused with idiopathic dilatation of the pulmonary artery, a corrected transposition of the great vessels, rheumatic or syphilitic heart disease with aortic insufficiency, and the Eisenmenger complex For a complete list see under Differential Diagnosis

Treatment is unsatisfactory but should be directed toward a reduction in the intrapulmonary pressure In the presence of cardiac failure digitalis is indicated

The prognosis is poor The pulmonary hypertension is usually progressive and eventually leads to cardiac failure and death

TREATMENT

At the present time treatment is only palliative Digitalis will help to strengthen the failing heart Any drugs[10] which offer the possibility of relaxation of the pulmonary vascular bed, such as priscoline or hexamethonium, may give some relief It is hoped that some drug may be discovered which will significantly reduce pulmonary pressure

The possibility of an allergic condition should be investigated As previously mentioned, the author has seen one patient who was greatly benefited by a summer in Maine, a part of the country which is known to be free from pollens and offers relief to many persons with hay fever Poppe, in discussion of a paper by Kaunitz and Andersen,[11] reported one patient in whom denervation of the lungs caused a long sustained reduction in the pulmonary pressure, but to the author's knowledge this is the only case in which denervation has been successful

There are probably many causes of "primary" pulmonary hypertension Nevertheless, until the etiology is known and a specific therapy is available, any measures which may reduce the pulmonary pressure should be given a trial

PROGNOSIS

The prognosis varies with the severity of the pulmonary hypertension It undoubtedly also varies with the etiology In very young patients in whom the pulmonary hypertension is more probably due to congenital abnormality of the pulmonary vascular bed, the slow expansion of the normal lung may compensate to a certain extent for the abnormality in the pulmonary vascular bed Consequently there may be considerable improvement during childhood A number of women have reached maturity and married and a few have been known to bear normal children Such, however, are the exception The condition generally progresses to an early fatal termination Death may result from cardiac failure or the circulation may become so sluggish that it is incompatible with life

SUMMARY

"Primary" pulmonary hypertension means that there is increased resistance in the pulmonary vascular bed and that the heart itself is normally formed

The etiology is obscure and it is probable that there is more than a single etiological factor

"Primary" pulmonary hypertension may date from birth and there may be no evidence of infection demonstrated either during life or at autopsy, hence it is believed to be caused by a congenital abnormality in the pulmonary vascular

bed The increased resistance in the pulmonary vascular bed increases the work of the right side of the heart, there results right ventricular hypertrophy and dilatation of the main pulmonary artery and its branches

The course of the circulation is normal, except when the foramen ovale is held open by the high pressure in the right auricle and a right to-left shunt is thereby established

Dyspnea is the outstanding complaint

Engorgement of the liver and ascites develop as cardiac failure becomes severe

Syncope and precordial pain are late manifestations

The heart undergoes progressive enlargement, which is mainly right sided The pulmonic second sound is accentuated and there may be a long, loud, early diastolic murmur

Fluoroscopy reveals a fullness of the pulmonary conus, there may be a con spicuous hilar dance but the periphery of the lungs is clear

The electrocardiogram shows marked right axis deviation and evidence of right ventricular hypertrophy

The diagnosis is based upon the finding of a right sided cardiac enlargement and a markedly accentuated second sound over the pulmonary area The size of the heart, the age at which the patient develops symptoms, and the rapidity with which these advance to cardiac failure depend upon the etiology and the severity of the pulmonary vascular changes

The diagnosis is definitely established by cardiac catheterization and the finding of a high pressure in the pulmonary artery, with a normal wedge pressure, and no evidence of intracardiac shunt.

The malformation must be differentiated from other conditions which cause right sided cardiac strain with or without persistent cyanosis In early infancy the condition may be confused with a tetralogy of Fallot or a complete transposition of the great vessels As the heart enlarges, its contour becomes similar to that of a pure pulmonary stenosis In the absence of cyanosis, the condition is most commonly confused with idiopathic dilatation of the pulmonary artery, a corrected transposition of the great vessels, rheumatic or syphilitic heart disease with aortic insufficiency, and the Eisenmenger complex For a complete list see under Differential Diagnosis

Treatment is unsatisfactory but should be directed toward a reduction in the intrapulmonary pressure In the presence of cardiac failure digitalis is indicated

The prognosis is poor The pulmonary hypertension is usually progressive and eventually leads to cardiac failure and death

TREATMENT

At the present time treatment is only palliative Digitalis will help to strengthen the failing heart Any drugs[10] which offer the possibility of relaxation of the pulmonary vascular bed, such as priscoline or hexamethonium, may give some relief It is hoped that some drug may be discovered which will significantly reduce pulmonary pressure

The possibility of an allergic condition should be investigated As previously mentioned, the author has seen one patient who was greatly benefited by a summer in Maine, a part of the country which is known to be free from pollens and offers relief to many persons with hay fever Poppe, in discussion of a paper by Kaunitz and Andersen,[11] reported one patient in whom denervation of the lungs caused a long sustained reduction in the pulmonary pressure, but to the author s knowledge this is the only case in which denervation has been successful

There are probably many causes of 'primary" pulmonary hypertension Nevertheless, until the etiology is known and a specific therapy is available, any measures which may reduce the pulmonary pressure should be given a trial

PROGNOSIS

The prognosis varies with the severity of the pulmonary hypertension It undoubtedly also varies with the etiology In very young patients in whom the pulmonary hypertension is more probably due to congenital abnormality of the pulmonary vascular bed, the slow expansion of the normal lung may compensate to a certain extent for the abnormality in the pulmonary vascular bed Consequently there may be considerable improvement during childhood A number of women have reached maturity and married and a few have been known to bear normal children Such, however, are the exception The condition generally progresses to an early fatal termination Death may result from cardiac failure or the circulation may become so sluggish that it is incompatible with life

SUMMARY

Primary" pulmonary hypertension means that there is increased resistance in the pulmonary vascular bed and that the heart itself is normally formed

The etiology is obscure and it is probable that there is more than a single etiological factor

'Primary' pulmonary hypertension may date from birth and there may be no evidence of infection demonstrated either during life or at autopsy, hence it is believed to be caused by a congenital abnormality in the pulmonary vascular

When the high pressure in the lesser circulation is due to changes in the pulmonary vascular bed and the heart itself is normally formed, the course of the circulation is as follows

The blood from the right auricle flows into the right ventricle and is pumped out into the pulmonary artery in the normal manner If the pressure in the pulmonary artery exceeds that in the descending aorta, a considerable volume of blood in the pulmonary artery will pass through the ductus arteriosus to the descending aorta The blood in the pulmonary artery which flows to the lungs is oxygenated in the lungs and returned in the normal manner to the left auricle, thence it flows into the left ventricle and is pumped out through the aorta to the systemic circulation Consequently the head and the upper extremities receive fully oxygenated blood from the left ventricle, whereas the trunk and lower extremities receive a mixture of fully oxygenated blood from the left ventricle and of venous blood which flows through the ductus arteriosus to the descending aorta It follows that the blood directed to the lower extremities is not fully oxygenated The blood from the upper extremities is returned by the superior vena cava to the right auricle and that from the lower extremities is returned by the inferior vena cava to the right auricle There the cycle starts again (see Diagram XVIII–3)

When the pulmonary hypertension is due to a severe malformation on the left side of the heart, the course of the circulation is altered only by the fact that a considerable part of the oxygenated blood returned to the left auricle will be shunted through the auricular defect into the right auricle, and consequently a mixture of oxygenated blood from the left auricle and venous blood from the right auricle will flow into the right ventricle and be pumped out through the pulmonary artery to the lungs and through the ductus arteriosus to the descending aorta Theoretically this means that the oxygen saturation of the blood in the descending aorta is not as low as when there is no shunt at the auricular level Nevertheless, inasmuch as the volume of the shunt varies with the severity of the pulmonary hypertension and the size of the ductus arteriosus, the oxygen unsaturation of the arterial blood in the descending aorta is subject to considerable variation Regardless of the presence or absence of an auricular defect and a malformation in the left side of the heart, the head and the upper extremities receive fully oxygenated blood which has been pumped from the left ventricle into the aorta and in both instances the trunk and lower extremities receive some venous blood (see Diagram XVIII–4)

B Pulmonary Hypertension with Persistent Patency of the Ductus Arteriosus

Pulmonary hypertension combined with persistent patency of the ductus arteriosus produces a distinctive clinical syndrome when the pressure in the pulmonary artery is greater than the systemic pressure

NATURE OF THE MALFORMATION

The essential feature of this malformation is the combination of severe pulmonary hypertension and persistent patency of the ductus arteriosus Consequently any condition which causes the pulmonary pressure to be higher than the systemic pressure, when it occurs in combination with persistent patency of the ductus arteriosus, will produce this clinical syndrome [1-13] The pulmonary hypertension may be primary, that is, due to increased resistance in the pulmonary vascular bed, or it may be secondary to a severe malformation on the left side of the heart combined with a gross defect in the auricular septum

The malformation may be that of severe mitral stenosis or there may be defective development of the left ventricle and marked hypoplasia of the ascending aorta In either instance, if there is also a gross defect in the auricular septum, as soon as the pressure in the left auricle exceeds that in the right auricle, a considerable volume of the blood returned to the left auricle will be shunted into the right auricle and thence to the right ventricle The mixture of oxygenated blood from the left auricle and of venous blood from the right auricle, which flows into the right ventricle, will be pumped out through the pulmonary artery to the lungs and through the ductus arteriosus to the descending aorta

It is conceivable, although still unproven, that an excessively large ductus arteriosus in which the pulmonary hypertension is initially compensatory may ultimately lead to irreversible changes in the lungs which will progress to such a point that the pressure in the pulmonary artery will exceed systemic pressure Consequently the flow of blood through the ductus arteriosus will again be reversed The author believes that this *may* occur when a child with an enormous ductus arteriosus regains compensation and is extremely active It is, however, the exception, not the rule

COURSE OF THE CIRCULATION

The course of the circulation varies with the nature of the pulmonary hypertension

Diagram XVIII–3

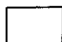
Primary pulmonary hypertension with patency of the ductus arteriosus

In this malformation there is increased resistance in the pulmonary vascular bed and the ductus arteriosus is patent

The blood from the right auricle flows into the right ventricle and is pumped into the pulmonary artery Part of the blood in the pulmonary artery flows to the lungs and, because of the high pressure in the pulmonary artery part flows through the ductus arteriosus into the descending aorta The blood which flows to the lungs is oxygenated and is returned in the normal fashion to the left auricle Thence it flows to the left ventricle and is pumped out through the aorta to the systemic circulation The blood which flows to the head and the upper extremities is returned in the normal manner by the superior vena cava to the right auricle The blood which flows through the arch of the aorta to the descending aorta meets the venous blood which flows through the ductus arteriosus to the descending aorta This mixture of arterial and venous blood flows to the trunk and the lower extremities and is returned by the inferior vena cava to the right auricle. There the cycle starts again

Clinical diagnosis is based upon the fact that the blood which flows to the lower extremities is not fully saturated hence the feet are cyanotic and the head and the upper extremities of normal color The heart is normal in size The pulmonic second sound is accentuated and there may be marked pulmonary insufficiency The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy

Diagram XVIII-3



Arterial blood (fully saturated)



Venous and arterial blood
Cyanosis visible



Small admixture of venous blood
No visible cyanosis



Venous blood

Diagram XVIII-4

Primary pulmonary hypertension combined with a severe left sided heart lesion and persistent patency of the ductus arteriosus

In this malformation there is a gross abnormality on the left side of the heart either mitral stenosis or underdevelopment of the left ventricle and hypoplasia of the ascending aorta, or some other severe left sided cardiac lesion combined with a gross defect in the auricular septum

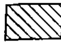
Under such circumstances the blood from the right auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs and also through the ductus arteriosus to the descending aorta The blood which flows to the lungs is fully oxygenated and is returned in the normal manner to the *left auricle*. Owing to the malformation on the left side of the heart, it is difficult for the blood in the left auricle to flow into the left ventricle and to be pumped out through the aorta to the systemic circulation. Nevertheless the blood which does flow to the left ventricle and thence to the systemic circulation is fully oxygenated The high pressure in the left auricle combined with the defect in the auricular septum, means that a considerable volume of oxygenated blood is shunted from the left auricle to the right auricle, where it mixes with the venous blood returned by the superior vena cava and the inferior vena cava to that chamber This mixture of oxygenated and venous blood flows into the right ventricle So the cycle continues

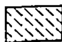
In this combination of anomalies two factors increase the pressure in the pulmonary artery The right ventricle pumps blood through the ductus arteriosus to the descending aorta hence the pressure in the pulmonary artery is the same as that in the descending aorta In addition the high pressure in the left auricle elevates the pressure in the pulmonary capillary bed Consequently there is severe pulmonary hypertension If the pressure in the descending aorta is significantly lower than that in the pulmonary artery as so frequently occurs with a severe left sided lesion some venous blood in the descending aorta may be forced back into the left subclavian artery, causing cyanosis of the left hand as well as of the feet.

Clinical diagnosis The difference in cyanosis between the two hands or between the hands and the feet, permits the diagnosis to be made at sight Although special studies may be necessary to determine the existence of a malformation on the left side of the heart, such a lesion should be suspected if the left hand is more cyanotic than the right hand

Diagram XVIII-4


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

artery may be so great that there is clubbing of the fingers of the left hand but not of the right Under such circumstances the superficial appearance is that of unilateral cyanosis namely, the left side of the body appears to be cyanotic and the right side of normal color Usually careful examination of the back will show that the head and neck and the tops of the shoulders are of normal color and [illegible] The line of demarcation of the cyanosis [illegible] part of the right shoulder receives its blood supply [illegible] tery, which carries fully oxygenated blood

The difference in cyanosis between the upper and lower extremities is usually sufficiently marked so that the diagnosis can be made at sight

Polycythemia develops slowly Initially the patient is not polycythemic and may even be anemic In the presence of anemia the volume of reduced hemoglobin circulating through the lower extremities may be too slight to produce 'visible' cyanosis As the polycythemia increases, the patient has the high color and the plethoric appearance so commonly seen in polycythemia vera

Difficulty in feeding and *failure to gain* are common complaints in early infancy

Repeated pulmonary infections are of such frequent occurrence that it sometimes seems as if the infant has hardly recovered from one infection before he develops another

Dyspnea on exertion is also a common complaint It varies with the severity of the pulmonary hypertension It is usually the development of exertional dyspnea which brings the patient to the doctor Thereafter dyspnea persists as the outstanding difficulty and becomes increasingly severe throughout the remainder of the patient's life

CARDIAC FINDINGS

The size of the heart varies with the underlying pathology If the condition is one of 'primary' pulmonary hypertension combined with patency of the ductus arteriosus the heart may be remarkably small, because the ductus arteriosus acts as an escape valve for the high pressure in the pulmonary artery (see Figure XVIII–4) Indeed the patency of the ductus arteriosus means that the pressure in the pulmonary artery cannot greatly exceed that in the systemic circulation and this in turn limits the work required of the heart Nevertheless, the right ventricle becomes hypertrophied and the pulmonary artery and its main branches are dilated consequently the pulmonary conus is prominent *The pul*

PHYSIOLOGY OF THE MALFORMATION

When the ductus arteriosus remains patent, as soon as the pressure in the pulmonary artery exceeds systemic pressure, the direction of the flow of blood *through the ductus will* be reversed, that is, blood will flow from the pulmonary artery into the descending aorta. For this reason the condition is frequently called a "reversed ductus." When this occurs, the ductus arteriosus acts as an escape valve, it prevents the pressure in the pulmonary artery from ever greatly exceeding systemic pressure and thereby lessens the load placed upon the heart. It does not, however, *limit* the extent of injury to the lungs. There is progressive narrowing of the pulmonary vascular bed which causes an ever increasing volume of blood to be shunted through the ductus arteriosus into the descending aorta.

If there is a severe lesion on the left side of the heart, the pressure in the systemic circulation is frequently abnormally low. Hence there is a greater tendency for the pressure in the pulmonary artery to be significantly higher than that in the descending aorta. There is a correspondingly greater likelihood for some of the blood which is shunted from the pulmonary artery through the ductus arteriosus to the descending aorta to be directed back into the left subclavian artery. Under such circumstances the left hand may appear slightly cyanotic. It follows that, when the left hand is more cyanotic than the right, the chances are in favor of the existence of a malformation on the left side of the heart.

CLINICAL FINDINGS

The distribution of the cyanosis gives the clue to the diagnosis. The head and the upper extremities are of normal color, whereas the lower extremities, which receive the mixture of oxygenated blood from the ascending aorta and of venous blood from the pulmonary artery, are cyanotic. When the pressure in the aorta is lower than that in the pulmonary artery, owing to the close proximity of the left subclavian artery to the point of entrance of the ductus arteriosus, some venous blood is forced back up the aorta and not infrequently the left subclavian artery receives some venous blood, together with the fully oxygenated blood from the ascending aorta. Consequently the left hand may be more cyanotic than the right. Often the difference in cyanosis between the two hands is not great, even the difference in cyanosis between the left hand and the feet may not be convincing but, upon comparison of the right hand with either foot, the difference in the color is readily apparent.

At times the oxygen unsaturation of the arterial blood in the left subclavian

X RAY AND FLUOROSCOPIC FINDINGS

When primary pulmonary hypertension *is associated* with persistent patency of the ductus arteriosus, the heart is usually normal in size but, when the condition is associated with a malformation on the left side of the heart, there may be considerable cardiac enlargement *In the anterior* posterior position the pulmonary conus is prominent and the main branches of the pulmonary artery are slightly dilated (see Figure XVIII-4) Sometimes there is enormous dilatation of the pulmonary artery and its main branches, as shown in Figure XVIII-5, in this instance the condition had progressed so far that there was calcification in the main branches of the pulmonary artery *In the left anterior*-oblique position the right ventricle usually appears to be slightly to moderately enlarged The right anterior-oblique position is usually best for the determination of left auricular enlargement If the left auricle is enlarged, it is strong evidence that the pulmonary hypertension is secondary to a serious abnormality of the left auricle or the left ventricle and that the defect in the auricular septum is small

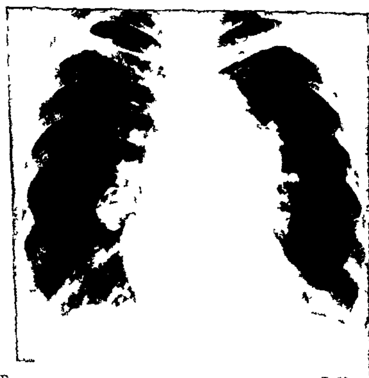


FIGURE XVIII-5 "Primary pulmonary hypertension and patency of the ductus arteriosus Adult

Note calcification in the aorta and the main branches of the pulmonary artery



FIGURE XVIII-4 Primary ' pulmonary hypertension and patency of the ductus arteriosus Adult

monic second sound is accentuated and usually reduplicated There may be a *long, loud early diastolic murmur* along the left sternal border due to pulmonary insufficiency

When the pulmonary hypertension is secondary to a malformation of the left auricle or the left ventricle, the heart may or may not be enlarged Furthermore, there may or may not be clinical evidence of mitral stenosis In some instances a presystolic murmur may be audible at the apex In other instances a severe malformation of the left side of the heart may produce no abnormal murmur or thrill A severe malformation on the left side of the heart frequently causes the pressure in the systemic circulation to be abnormally low

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FIGURE XVIII–5 Primary pulmonary hypertension and patency of the ductus arteriosus Adult

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Cardiac catheterization will reveal high pressure in the right ventricle and in the pulmonary artery Since the direction of the flow of blood is through the ductus arteriosus to the descending aorta, it is frequently possible to pass the catheter into the descending aorta Cardiac catheterization, however, gives little or no information concerning the presence or absence of a malformation on the left side of the heart

Angiocardiography is less dangerous in this combination of anomalies than when the ductus arteriosus is closed, as the ductus arteriosus acts as an escape valve and thus prevents the accumulation of dye in the lungs, furthermore, the brain is supplied with fully oxygenated blood

Angiocardiography will reveal that the descending aorta fills simultaneously with the pulmonary artery In patients with mitral stenosis or marked hypoplasia of the mitral valve, the dye will linger for a long time in the left auricle, the left ventricle and ascending aorta may never be visualized

Aortography may be helpful in the determination of the size of the ascending aorta and the exclusion of a coarctation of the aorta In order to obtain a satisfactory aortogram, the dye must be injected into an artery on the same side of the body as that toward which the aorta arches that is, with a normal left aortic arch the test should be performed through the left brachial artery If the dye is injected on the opposite side to that of the descending aorta, the increased pressure transmitted from the pulmonary artery to the descending aorta will lessen the flow of blood from the ascending aorta to the descending aorta, consequently the dye may pass only up the carotid arteries to the head This is especially prone to occur if the left hand is cyanotic Under such circumstances the aortogram may simulate that of a complete interruption of the aortic arch even though in reality the arch is intact

When aortography is properly performed, it is of great diagnostic aid The arch of the aorta and the ductus arteriosus may both be visualized The contrast media enters the ductus and delineates it but, owing to the high pressure in the pulmonary artery, no dye enters the lungs This finding is in striking contrast to the simultaneous visualization of the aorta and the pulmonary arteries in an infant with a large ductus arteriosus and pulmonary hypertension when the predominant shunt is from the aorta to the lungs (see Chapter xx)

DIAGNOSIS

The diagnosis may be made at sight when the left hand is cyanotic and the right hand is of normal color The diagnosis can also be made at sight when the face and the upper extremities are of normal color and the feet are cyanotic

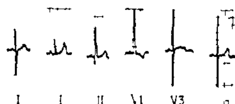


Figure xviii–6 Primary pulmonary hypertension and patency of the ductus arteriosus

Fluoroscopy frequently reveals pulsations in the main pulmonary artery but, owing to the fact that the ductus arteriosus acts as an escape valve, these pulsations are generally minimal and do not extend into the small branches of the pulmonary artery The periphery of the lungs is usually abnormally clear

ELECTROCARDIOGRAPHIC FINDINGS

The standard leads show a right axis deviation and the unipolar precordial leads show evidence of marked right ventricular hypertrophy (see Figure xviii–6) Inasmuch as the patency of the ductus arteriosus relieves the strain on the right ventricle, the evidence of right ventricular hypertrophy is frequently not as marked as when primary pulmonary hypertension occurs as an isolated malformation Indeed, the author has seen one patient in whom the lesion on the left side of the heart was such as to give a left axis deviation, but even in that instance the precordial leads showed evidence of right ventricular hypertrophy

SPECIAL TESTS

A difference in the oxygen saturation of arterial blood between the right brachial artery and the femoral artery is characteristic of this malformation, because the head and the upper extremities receive fully oxygenated blood from the ascending aorta and the femoral artery receives some venous blood which is directed through the ductus arteriosus to the descending aorta In order to be certain that the difference between the right brachial artery and the femoral artery is significant, the two blood samples must be drawn simultaneously It is, of course, unfortunate but inevitable that the sample of blood from the femoral artery will approximate that of normal venous blood If, however, a sample of venous blood is drawn, the oxygen content of the venous blood will be even lower than that of the femoral arterial blood Furthermore, the pressure in the femoral artery is always higher than that in the femoral vein

Additional studies are usually necessary to determine whether the pulmonary hypertension is primary or secondary, that is, whether the principal difficulty occurs in the lungs or in the heart Such knowledge is obviously essential as a guide to therapy

ondary *to* the large ductus arteriosus or whether the patency of the ductus arteriosus is secondary to the pulmonary hypertension

Pulmonary hypertension secondary to the increased circulation to the lungs is the rule in infants with a large ductus arteriosus Such *infants* show great cardiac enlargement, a systolic and a mid-diastolic murmur over the precordium, enlargement of the left auricle, increased vascular shadows, and electrocardiographic evidence of 'combined hypertrophy

In contrast to these findings, when the pulmonary hypertension is primary and the ductus arteriosus has remained patent, the patient shows cyanosis of the lower extremities, the heart is usually of normal size, there may or may not be a systolic murmur, the pulmonary vascularity is reduced, and the electrocardiogram shows evidence of right ventricular hypertrophy

An aortogram sharply differentiates the two conditions When the pulmonary hypertension is secondary to the flow of blood through the ductus arteriosus, dye is seen in the lungs as the aorta is visualized, whereas when the pulmonary hypertension is primary, although the ductus arteriosus may be opacified, no dye enters the lungs

Cardiac catheterization will aid in the differentiation of the two conditions When the pulmonary hypertension is secondary to a large ductus, it is the large left to-right shunt which causes the hypertension, under such circumstances there is a marked increase in the oxygen content of the blood in the pulmonary artery On the other hand, when the pulmonary hypertension is primary, the shunt is from right to-left, there is little or no evidence of a left to-right shunt, and no increase in the oxygen content of the blood in the pulmonary artery

Thus the extreme of the one is readily differentiated from the extreme of the other In early infancy the two conditions are usually readily differentiated In older patients there may be real difficulty in the differentiation of pulmonary hypertension combined with persistent patency of the ductus arteriosus from a patent ductus arteriosus which causes pulmonary *hypertension* (see Chapter xx)

If the flow of blood from the pulmonary artery *to the aorta is sufficient to* cause cyanosis of the lower extremities, or even if there is a clearly detectable difference in the oxygen saturation of the arterial blood in the brachial and femoral arteries the pulmonary hypertension has *progressed* to the point where it, not the ductus arteriosus, is the primary difficulty *Under* such circumstances, unless the pressure in the pulmonary artery can be relieved, ligation of the ductus arteriosus is usually contraindicated

Complete transposition of the great vessels combined with a patency of the

Sometimes the history of a slight cardiac disability throughout life, combined with the finding of a loud *diastolic* murmur, will direct attention to the possibility of pulmonary hypertension and will lead to careful examination of the color of the right hand and of the feet

The determination of the oxygen saturation of the arterial blood samples drawn simultaneously from the right arm and from either of the femoral arteries clinches the diagnosis

Unless there is frank evidence of mitral stenosis or evidence of left auricular enlargement, further studies are usually necessary to determine whether the pulmonary hypertension is primary or secondary to a severe malformation on the left side of the heart

DIFFERENTIAL DIAGNOSIS

This condition requires differentiation from 'primary" pulmonary hypertension (see Section A) and also from other conditions which cause a difference in cyanosis between the upper and the lower extremities, such as coarctation of the aorta with the ductus arteriosus opening below the constriction, patency of the ductus arteriosus associated with pulmonary hypertension, complete transposition of the great vessels, and complete interruption of the isthmus of the aorta when the descending aorta is continuous with the pulmonary artery through the ductus arteriosus

Coarctation of the aorta with the ductus arteriosus opening below the constriction theoretically would give the same clinical picture as a pulmonary hypertension with a persistent patency of the ductus arteriosus An infant with a coarctation of the aorta frequently develops severe right sided cardiac failure, furthermore, as the ductus periodically clamps down, no pulse is palpable in the femoral artery Aortography performed through the left subclavian artery will readily show whether or not there is a coarctation of the aorta

If there is cyanosis of the left hand, it is clear evidence that there is reflux of blood from the pulmonary artery into the left subclavian artery, hence there cannot be a constriction of the aorta between the left subclavian artery and the point of entrance of the ductus arteriosus

Occasionally a large patent ductus arteriosus presses upon and constricts the aorta and thus causes a functional coarctation of the aorta An aortogram usually reveals the true nature of the difficulty

A large patent ductus arteriosus associated with pulmonary hypertension presents a diagnostic problem which is both extremely difficult and important Almost invariably the question arises whether the pulmonary hypertension is sec

Coarctation of the aorta can be corrected by surgery In young infants who suffer from severe cardiac failure, it is, however, usually preferable to close the ductus and at a later date to correct the coarctation (see Chapter xxvii)

If the pulmonary hypertension is secondary to severe mitral stenosis, early relief of the mitral stenosis *may* be of benefit When the mitral stenosis is due to rheumatic fever, operation is relatively simple, but when the mitral stenosis is congenital, the problem is much more difficult (see Chapter xxx) All too frequently the mitral valve and the left auricle may be so grossly abnormal that it is impossible to improve the circulation Dr Clifford Parsons[14] has reported one successful operation on a congenital mitral valve, but in this case the operation did not alter the pulmonary pressure

Complete interruption of the isthmus of the aorta offers a challenge to the surgeon to reunite the proximal and distal ends of the aorta, either directly or by graft, and then to divide the ductus arteriosus Unfortunately, however, the condition is usually associated with additional cardiac abnormalities which must also be corrected in order to help the infant

PROGNOSIS

The prognosis varies with the etiology and the severity of the pulmonary hypertension By and large the prognosis is better when 'primary" pulmonary hypertension is combined with patency of the ductus arteriosus than when the ductus undergoes normal obliteration This is an additional reason for the author s great hesitancy in recommending surgical closure of the ductus unless the ductus arteriosus is clearly the cause of the hypertension The author has known several women in their early twenties who were only slightly incapacitated by this condition, one woman has married and has had one normal pregnancy without great cardiac difficulty Thus, although the condition is extremely serious, it may be compatible with life for a number of years

SUMMARY

Pulmonary hypertension combined with persistent patency of the ductus arteriosus produces a distinctive clinical syndrome

The pulmonary hypertension may be primary or may be secondary to a severe malformation on the left side of the heart In either case, owing to the high pressure in the lungs, blood flows from the pulmonary artery through the ductus arteriosus to the descending aorta Consequently the head and the upper extremities receive fully oxygenated blood and the trunk and the lower extremities receive a mixture of venous and arterial blood Therefore the head and

ductus arteriosus may be confused with the malformation under discussion because of the difference in cyanosis between the upper and the lower extremities The difference in cyanosis in a complete transposition of the great vessels is the reverse of that which occurs in primary pulmonary hypertension and persistent patency of the ductus arteriosus that is, the head and the upper extremities are more cyanotic than are the trunk and the lower extremities Moreover, when there is a complete transposition of the great vessels, inasmuch as the pressure in the aorta is normal and that in the pulmonary artery is so high that blood flows from the pulmonary artery into the descending aorta, the line of demarcation of the cyanosis lies at the brim of the pelvis In contrast to this, when the pressure in the systemic circulation is so low that the systemic pressure is maintained by the flow of blood from the pulmonary artery through the ductus arteriosus to the aorta, the line of demarcation of the cyanosis lies high up on the shoulder girdle

Complete interruption of the isthmus of the aorta may also be confused with this malformation An aortogram, properly taken, should enable the physician to make the correct diagnosis

TREATMENT

The treatment obviously depends upon the cause of the pulmonary hypertension

If the hypertension is secondary to a left sided cardiac lesion, attention should be directed to the correction of the cardiac anomaly

The crucial question is whether the pulmonary hypertension is secondary to the patency of the ductus arteriosus or the pulmonary hypertension is primary and the ductus arteriosus acts as an escape valve to relieve the high pressure in the lungs

The differentiation of these two conditions is extremely important because, if the ductus arteriosus is the cause of the pulmonary hypertension, ligation of the ductus restores the heart and circulation to normal (see Chapter xx), whereas, if the pulmonary hypertension is primary, ligation of the ductus arteriosus usually causes great dilatation of the right side of the heart and may lead to cessation of heart action Even in instances in which the patient survives the closure of the ductus, the benefit derived therefrom is doubtful Indeed, closure of the ductus arteriosus may ultimately do harm because it closes the escape valve It is for these reasons that the author is strongly opposed to closure of the ductus arteriosus in a patient with severe pulmonary hypertension unless the pulmonary pressure can be reduced

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the upper extremities are of normal color and the trunk and the lower extremities are cyanotic The distribution of the cyanosis permits diagnosis at sight

The cardiac findings vary with the etiology of the hypertension If the pulmonary hypertension is primary, the heart is usually relatively small The second sound over the pulmonary area is accentuated and frequently there is a loud diastolic murmur audible along the left sternal border

X ray and fluoroscopy show that the heart is not greatly enlarged, there may be a minimal hilar dance The periphery of the lungs is abnormally clear If the pulmonary hypertension is secondary to a malformation of the left auricle or the left ventricle, x ray and fluoroscopy may reveal evidence of left auricular enlargement

The electrocardiogram usually shows right axis deviation and evidence of right ventricular hypertrophy

The determination of the oxygen saturation of samples of arterial blood taken simultaneously from the right brachial artery and either femoral artery will clinch the diagnosis

Although diagnosis may be made at sight, special studies are usually necessary to determine the nature of the pulmonary hypertension

Angiocardiography is of greater aid than cardiac catheterization It demonstrates the flow of blood from the pulmonary artery to the descending aorta and it also gives some information concerning the left side of the heart

The condition requires differentiation from coarctation of the aorta combined with patency of the ductus arteriosus, from simple patency of a large ductus arteriosus in which there is pulmonary hypertension, and also from complete interruption of the isthmus of the aorta in which the descending aorta is continuous with the pulmonary artery through the ductus arteriosus

The treatment varies with the etiology

Prognosis is guarded but the condition may be compatible with life for a number of years

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NATURE OF THE MALFORMATION

The essential feature of the malformation concerns the anomaly of the tricuspid valve and of the upper portion of the wall of the right ventricle One or more leaflets of the tricuspid valve are fused with the ventricular endocardium over large areas Consequently it is often difficult to identify the individual leaflets with certainty The anterior leaflet is the least involved, indeed, it is often normal The medial or septal leaflet may or may not be normal Frequently it is sealed to the septal wall and in some instances it is entirely absent The posterior leaflet is usually the most malformed and is displaced down toward the apex of the right ventricle In most instances the upper portions of the leaflet are blended with the endocardium of the right ventricle and the distal margins of the valve which are free become fused together to form a basket network deep within the right ventricle Consequently there is no valve at the normal tricuspid orifice but a new valve is formed by the basket network Sometimes this new valve has a *normal orifice* which opens into the lower portion of the right ventricle In other instances there is no true orifice and the blood passes through the interstices of the basket network into the outflow tract of the right ventricle In either event the new valve which is formed deep within the right ventricle divides this chamber into two parts That portion of the right ventricle which *lies above the valve* becomes incorporated into the right auricle and only that portion of the right ventricle which lies below the valve is of functional importance in the expulsion of blood to the lungs

The extent to which the tricuspid valve is plastered against the right ventricle and the position of the new valve vary from case to case In some instances the abnormality is relatively slight and the position of the new valve is such that the lower chamber is the larger of the two cavities Such is the anomaly shown in Figure xix–1 In other instances virtually the entire tricuspid valve is plastered against the endocardium of the right ventricle and the new valve extends nearly to the apex of the right ventricle Under such circumstances the lower chamber, which is the outflow tract of the right ventricle, is abnormally small, as illustrated in Figures xix–2 and 3 Nevertheless, the tricuspid valve may be abnormally large and during ventricular systole may balloon upward into the right auricle, thus increasing the size of the lower chamber This chamber is, however, usually too small to receive all the blood which has been returned to the right auricle and to pump the blood through the pulmonary artery to the lungs

Engle and her associates[3] have pointed out that the extreme thinness of the

CHAPTER XIX

EBSTEIN'S ANOMALY

CONGENITAL DOWNWARD DISPLACEMENT OF THE TRICUSPID VALVE

IN 1866 Ebstein[1] first reported the malformation which now bears his name The anomaly consists of a congenital downward displacement of the tricuspid valve into the right ventricle The tricuspid valve is distorted in such a manner that one or more of its leaflets become plastered against the wall of the right ventricle, consequently there is no valve at the annulus fibrosus which separates the right auricle from the right ventricle For this reason Ebstein described the abnormality as a severe degree of congenital insufficiency of the tricuspid valve Actually in most instances a new valve is formed deep within the right ventricle, usually this valve is functionally sufficient The right ventricular wall above the anomalous valve is extremely thin The anomaly has been reviewed by Yater and Shapiro[2] and the clinical syndrome has been clarified by Engle et al [3] and by Schiebler et al [4]

EMBRYOLOGY

A brief review of the early embryology of the musculature of the heart will aid in understanding the nature of the malformation Streeter[5] has shown that the myocardium arises from a specialized portion of the visceral celomic wall and is separated from the endocardium of the primitive heart by the myoendocardial space which is filled with a homogenous transparent material known as the "cardiac jelly" As the myocardium develops, this jelly gradually disappears, except at strategic sites, where it persists to form the so-called endocardial cushions which are the precursors of the valves Thus, as Engle et al [3] have postulated, "any defect in the visceral celomic wall in the region where the right ventricle is to develop could not only cause defective development of the right ventricular myocardium but also by distortion of the position of the persisting primitive myoendocardial space could cause a malformation of the tricuspid valve" Be that as it may, defective development of the wall of the right ventricle above the anomalous tricuspid valve appears to be an integral part of the malformation

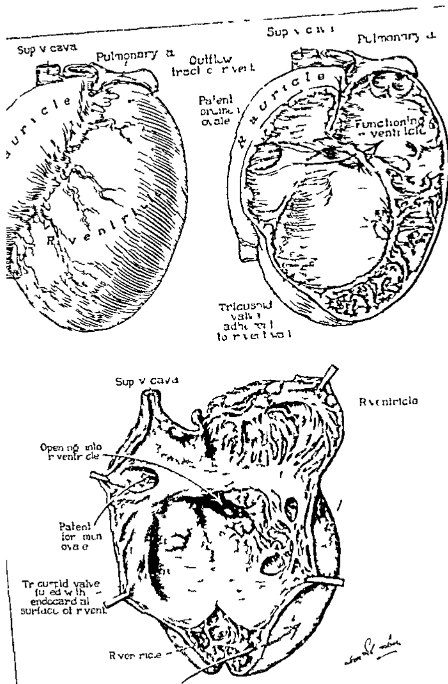


FIGURE XIX-2 Ebstein s anomaly with marked displacement of the tricuspid valve (same patient as in Figures XIX-5 7) Child

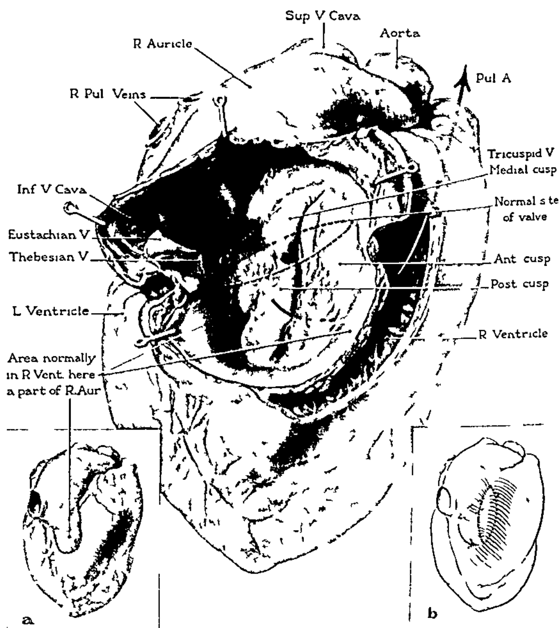


FIGURE XIX-1 Ebstein's anomaly with relatively slight displacement of the tricuspid valve (same patient as in Figure XIX-4) Young adult

myocardium in the upper portion of the right ventricular wall which lies above the new valve is an integral part of the malformation The myocardium in this region may be only 1 or 2 mm in thickness Localized aneurysmal dilatation of the right ventricular wall in this area has been reported by Blackhall Morison[6] and Brekke[7] In each instance the aneurysm was believed to be due to congenital weakness of the wall That portion of the wall of the right ventricle which lies below the valve is of normal thickness but the lower cavity is abnormally small, the ventricular septum is intact

In addition to the malformation of the tricuspid valve, there is frequently some abnormality either of the eustachian valves or of the thebesian valves Such anomalies are, however, only of academic interest

The foramen ovale may or may not be patent Although in some instances the foramen ovale closes in the normal manner and becomes completely sealed, it remains anatomically and functionally patent in two-thirds of the patients with this malformation If the valve covering the foramen ovale is *not* completely sealed, the high pressure in the right auricle will force the valve to open Indeed, the increased pressure in the right auricle caused by the misplaced tricuspid valve probably explains the high incidence of patency of the foramen ovale which occurs in association with this malformation[3] When the foramen ovale remains patent it acts as an escape valve for the high pressure in the right auricle Thereby it becomes of functional importance and a right to-left shunt is established

COURSE OF THE CIRCULATION

The course of the circulation depends upon whether or not the valve covering the foramen ovale becomes completely sealed So long as the foramen ovale is closed and in all instances in which the foramen ovale becomes completely sealed the circulation of the blood is normal Under such circumstances the blood from the right auricle flows into the right ventricle and is pumped out into the pulmonary artery The blood in the pulmonary artery is directed to the lungs where it is oxygenated, and is returned in the normal fashion by the pulmonary veins to the left auricle and thence to the left ventricle The blood in the left ventricle is pumped out by way of the aorta to the systemic circulation and is returned in the normal manner by the superior vena cava and the inferior vena cava to the right auricle (see Diagram XIX–1) Inasmuch as there is no communication between the two sides of the heart, there is no venous-arterial shunt Hence there is no cyanosis and no clubbing

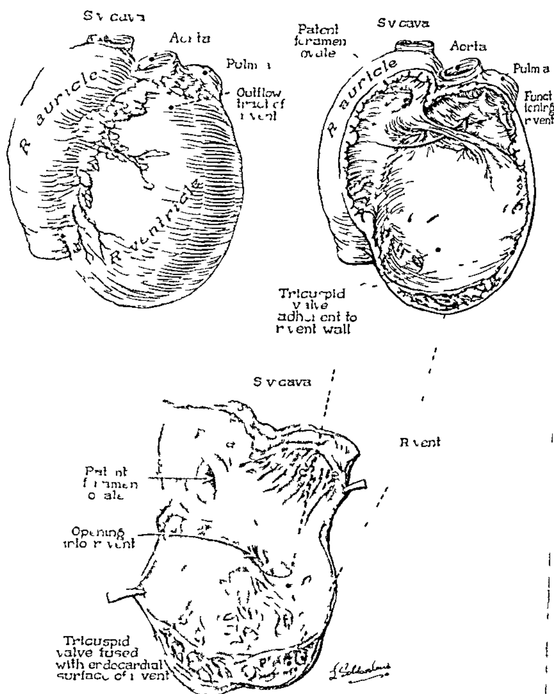


FIGURE XIX–5 Ebstein's anomaly with extreme displacement of the tricuspid valve (same patient as in Figure XIX–6) Young child

DIAGRAM XIX-1

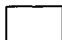
Ebstein's anomaly of the tricuspid valve

The essential feature of this malformation is a congenital downward displacement of the tricuspid valve into the right ventricle. Consequently the tricuspid valve bisects the right ventricle and part of the right ventricle is incorporated into the right auricle, the valve itself is usually competent.

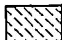
When the foramen ovale is completely closed, the blood from the right auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs where it is oxygenated The oxygenated blood is returned by the pulmonary veins to the left auricle Thence it flows to the left ventricle and is pumped out by way of the aorta to the systemic circulation It is returned in the normal fashion by the superior and inferior venae cavae to the right auricle. There the cycle starts again

Clinical diagnosis The clinical findings vary with the severity of the abnormality of the tricuspid valve If the displacement of the tricuspid valve is slight symptoms are minimal Weakness and fatigue are the outstanding complaints If there is great displacement of the tricuspid valve the malformation leads to great cardiac enlargement The heart sounds are weak Murmurs are variable there may be a systolic murmur only or there may be a to-and fro murmur over the precordium Cardiac arrhythmias are common The electrocardiogram usually shows low voltage complexes of long duration in V_1 and V and normal deflection over the left precordium The liver becomes engorged but does not pulsate Tricuspid insufficiency is a late manifestation Terminally there may also be edema of the extremities Sudden death is not uncommon

Diagram XIX–1


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

CLINICAL FINDINGS

The clinical findings vary with the severity of the abnormality and the extent of the displacement of the tricuspid valve. If the valvular abnormality is slight, the symptoms are minimal or absent and the condition is compatible with an active life. The greater the displacement of the tricuspid valve into the right ventricle, the more inefficient is the action of the right side of the heart and the earlier and the more severe are the symptoms.

Fatigue is more prominent than dyspnea. Even though the patient may show *persistent cyanosis* and tire easily, there is little dyspnea. The child is exhausted and *rests* but he is not short of breath. He does not squat when tired.

Palpitation is a common complaint, owing to the frequency of cardiac arrhythmias. Many patients suffer from numerous extrasystoles or repeated attacks of paroxysmal tachycardia. For this reason, in a person with an otherwise normal heart the persistence of a cardiac arrhythmia over a period of years should suggest the possibility of Ebstein's anomaly.

The presence or absence of cyanosis depends upon *the structure of the foramen ovale*. If, as previously mentioned, the valve covering the foramen ovale remains patent, the valve will be forced open by the increased pressure in the right auricle and a right-to-left shunt will be established. When a sufficient amount of reduced hemoglobin is shunted into the systemic circulation, the patient will develop persistent cyanosis. The wider the patency of the foramen ovale, the greater is the shunt and the earlier is the development of cyanosis. Although cyanosis may date from infancy, the development of cyanosis between two and five years of age is the rule rather than the exception.

Polycythemia develops after cyanosis makes its appearance. The red blood cell count, the amount of available hemoglobin, and the hematocrit reading gradually increase as the oxygen unsaturation of the arterial blood increases.

The liver is enlarged and congested and frequently may extend to the umbilicus. The force of the heart action is, however, too weak and the alternations of the auricular and ventricular contractions which occur in the auricle and auricularized portion of the right ventricle are too ineffective to cause pulsations at the margin of the liver.

CARDIAC FINDINGS

The heart may or may not be enlarged. In the initial stage *the right auricle* is increased in size at the expense of the right ventricle. Consequently the over-all size of the heart is normal.

If the tricuspid valve is markedly displaced into the right ventricle, the right auricle becomes a huge, ineffective chamber Since the right ventricle is not sufficiently large to receive all the blood from the right auricle, there is great dilatation of the right auricle and the pressure in that chamber gradually rises When the pressure in the right auricle exceeds that in the left auricle, if the valve covering the foramen ovale is not completely sealed, it will be forced open and a right to-left shunt will be established As soon as the volume of reduced hemoglobin so shunted to the systemic circulation becomes sufficiently great, the patient develops cyanosis The course of the circulation is shown in Diagram XIV-2

PHYSIOLOGY OF THE MALFORMATION

The altered structure of the heart mainly affects the manner in which the heart functions The abnormal position of the tricuspid valve renders it difficult for the right auricle to expel its blood into the functional portion of the right ventricle Although normally the auricles contract before the ventricles, in Ebstein's anomaly the right auricle does not contract in the normal manner because part of the right ventricle lies above the tricuspid valve Consequently the upper or genuine auricular portion of the right auricle contracts before its lower portion, which is composed of ventricular muscle Thus with each auricular contraction, the blood in the upper part of the auricle is directed through the tricuspid valve to the lower chamber, but with each ventricular contraction, when the tricuspid valve is closed the blood in the lower part of the auricle cannot be directed into the functioning part of the right ventricle and is merely pushed back into the upper portion of the auricle This mechanism causes the right auricle to be extremely ineffective in directing blood to the functioning portion of the right ventricle Thus, with each heart beat, the lower part of the right ventricle receives less than its normal quota of blood It follows that with each ventricular systole only a small quantity of blood is pumped out into the pulmonary artery The pressure in the right ventricle is, however, normal and the pressure in the pulmonary artery is also normal It is the abnormal position of the tricuspid valve which causes the right side of the heart to be extremely inefficient The condition leads to progressive dilatation and hypertrophy of the right auricle and the right ventricle The dilatation is far greater than the hypertrophy, the right side of the heart becomes enormously enlarged and the pressure in the right auricle gradually rises As previously stated, the valve covering the foramen ovale may be forced open and a right to left shunt thereby established

DIAGRAM XIX-2

Ebstein's anomaly of the tricuspid valve and patency of the foramen ovale

The essential feature of this malformation is a congenital downward displacement of the tricuspid valve into the right ventricle. The result is that the tricuspid valve bisects the right ventricle and part of the right ventricle is incorporated into the right auricle. The tricuspid valve is usually competent.

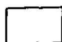
The abnormal position of the tricuspid valve causes the right auricle to be greatly enlarged and the smaller lower portion of the right ventricle is the only part of that chamber which is of functional importance The greater the displacement of the tricuspid valve, the larger is the right auricle and the less effective is that chamber in the direction of blood to the functional part of the right ventricle.

As the pressure in the right auricle rises, if the valve which covers the foramen ovale is not completely sealed, it will be forced open by the high pressure in the right auricle and a right to-left shunt will be established

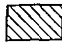
Some blood from the right auricle flows into the left auricle and the remainder of the blood in the right auricle flows into the lower part of the right ventricle and is pumped slowly to the lungs where it is oxygenated The oxygenated blood is returned in the normal fashion to the left auricle, where it meets the blood which has been shunted from the right auricle into that chamber This mixture of oxygenated and venous blood flows into the left ventricle and is pumped out through the aorta to the systemic circulation and returned by the superior vena cava and the inferior vena cava to the right auricle. There the cycle starts again

Clinical diagnosis The greater the displacement of the tricuspid valve the larger is the heart and the more probable it is that cyanosis is present. The patency of the foramen ovale acts as an escape valve for the high pressure in the right auricle but it also diverts the blood from the lungs and reduces the pulmonary blood flow Hence the lungs are exceptionally clear Weakness and fatigue are the outstanding complaints The heart is enlarged and often there is a to and fro murmur over the precordium and a gallop rhythm The liver is enlarged but does not pulsate The electrocardiogram shows low voltage curves of long duration in V_1 and V_2 and normal voltage in V_5 and V_6 Terminally there may be edema of the extremities Sudden death is not uncommon

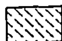
DIAGRAM XIX–2



Arterial blood (fully saturated)



Venous and arterial blood
Cyanosis visible



Small admixture of venous blood
No visible cyanosis



Venous blood

sternum, consequently there may *be nothing* characteristic in the size or shape of the heart, as illustrated in Figure XIX-4

Eventually (as shown in Figures XIX-5 and 6) the heart may be enormously enlarged both to the right and to the left The right auricle and the right ventricle may be so huge that they entirely obscure the left auricle and *the left ventricle* Even though the pulmonary arteries are of normal size, they are hidden behind the tremendous heart Because of the difficulty in *the direction of the blood to the lungs*, the lungs appear abnormally clear The hilar shadows are minimal and do not show expansile pulsations *Whenever the foramen ovale is not completely sealed* and the blood is shunted from the right auricle *to the left* auricle, the pulmonary blood flow is reduced Under such circumstances the lungs may appear so clear that the diagnosis of pulmonary stenosis may be erroneously entertained

Viewed in the left anterior-oblique position the heart may be flattened against the anterior chest wall for several inches, as shown in Figure XIX-5, and even when the patient is rotated at an angle of 60 degrees the left ventricle may

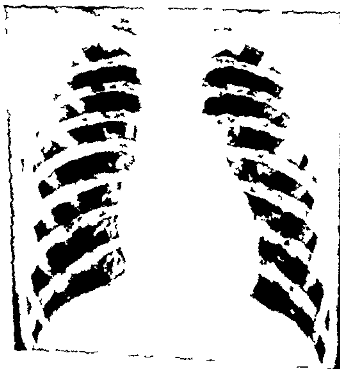


FIGURE XIX-4 Ebstein's anomaly with relatively slight displacement of the tricuspid valve (same patient as in Figure XIX-1) Young adult

The malformation leads to progressive cardiac enlargement The rate of cardiac enlargement varies with the severity of the abnormality of the tricuspid valve and the relative size of the two portions of the right ventricle The incorporation of the upper portion of the right ventricle into the right auricular chamber increases the size of this chamber but decreases the size of the functional portion of the right ventricle Furthermore, although the upper part of the right ventricle becomes incorporated into the right auricle, it does not contract simultaneously with the right auricle Thus first one part of the chamber contracts and then the other Only the upper portion of the right auricle contracts with auricular systole, the lower portion, which is composed of the 'auricularized' part of the right ventricle, contracts with ventricular systole Furthermore, it is only with auricular systole that the blood flows through the tricuspid valve into the functioning portion of the right ventricle The tricuspid valve closes with ventricular systole Consequently the contraction of the lower part of the right auricle, which occurs with ventricular systole, cannot direct the blood to the lower chamber Thus the action of the right auricle is extremely inefficient, the anomaly leads to enormous dilatation of the right auricle and the auricularized portion of the right ventricle The cardiac enlargement extends both to the right and to the left of the sternum and eventually the heart occupies most of the thoracic cavity

The heart sounds are weak and of poor quality There is often a gallop rhythm *Murmurs* are variable A *systolic murmur* is the rule There may be a *diastolic murmur,* and not infrequently there is a *to and fro murmur* which may simulate a pericardial friction rub Indeed, it is the persistence of such a murmur over a long period of time, and its occurrence in the absence of other signs or symptoms of infection, which help to differentiate this murmur from a true pericardial friction rub

Cardiac failure develops insidiously and may become severe It is mainly right sided heart failure with engorgement of the liver and edema of the extremities Although tricuspid insufficiency has been considered so common in Ebstein's anomaly as to be almost synonymous with it, actually, except as a terminal event, its occurrence is rare

X RAY AND FLUOROSCOPIC FINDINGS

In the early stages, or when the displacement of the tricuspid valve is slight, x ray and fluoroscopic findings may be normal The portion of the right ventricle which becomes incorporated into the right auricle is hidden behind the



FIGURE XIX-6 Ebstein's anomaly with extreme displacement of the tricuspid valve (same patient as in Figure XIX-3) Young child

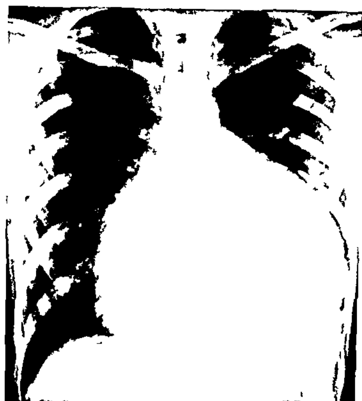
overlap the spine Under such circumstances the enlargement seen posteriorly is not due to a huge left ventricle but is caused by the backward displacement of the left ventricle by the huge right auricle and the right ventricle

Viewed in the right anterior-oblique position the heart appears to fill the entire chest cavity There is, however, no evidence of left auricular enlargement In both the anterior posterior position and the right anterior-oblique position, the esophagram is normal

The great enlargement of the heart and the weak pulsations cause the fluoroscopic appearance to simulate a pericardial effusion

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram, especially in older children and adults, may be a great aid in diagnosis Van Lingen and Bauersfeld[8] have shown that there is prolongation of both the auricular and the ventricular contractions The P waves are usually notched and of such long duration as to *increase* the P R interval Evidence of an intraventricular conduction disturbance is frequently seen in the standard leads The unipolar precordial leads give the clue to diagnosis The deflections in V_1 are of extremely low amplitude and of long duration The



Anterior posterior position



Left anterior-oblique position

FIGURE XIX–5 Ebstein's anomaly with marked displacement of the tricuspid valve (same patient as in Figure XIX–2) Child

whereas that in the right ventricle is normal The enormous size of the right auricle renders it difficult to pass the catheter *into the right* ventricle Moreover, catheterization may be extremely dangerous The auricularized portion of the right ventricle is so thin that there is danger that the catheter may pierce the wall In addition, the catheter may become entangled in the basket network of the tricuspid valve A further danger exists because of the susceptibility of patients with Ebstein s anomaly to cardiac arrhythmias In normal persons the manipulation of the catheter in the region of the tricuspid valve frequently produces arrhythmias Therefore it is not surprising that in patients with this malformation cardiac catheterization may initiate ventricular fibrillation or some other serious arrhythmia In the presence of tremendous cardiac dilatation prolonged auricular paroxysmal tachycardia may be fatal In addition, there is always the danger of cardiac arrest Therefore cardiac catheterization should not be undertaken lightly Nevertheless, Hernandez et al [9] have shown that intracardiac electrocardiograms taken during cardiac catheterization may be of great aid in diagnosis Normally the character of the electrocardiogram changes when the ventricle is entered In Ebstein s anomaly a ventricular type electrocardiogram may be obtained while the pressure curves indicate that the catheter is still in the auricle

Angiocardiography reveals prompt filling of the enormous right auricle and of the auricularized portion of the right ventricle, subsequently, as the lower portion of the right ventricle is filled, an even larger area becomes opacified (see Figure XIX-8) It is usually possible to see that the dye extends nearly to the margin of the cardiac silhouette, thus demonstrating the thinness of the right ventricular wall The dye lingers for a long time in the right auricle and only a small amount of blood and of contrast material is ejected into the lungs with each ventricular systole Consequently the lung fields are never well visualized The aorta may or may not be delineated *It is never filled until after the left auricle has* been visualized When there is a right to-left shunt through the foramen ovale, there may be faint early visualization of the left auricle, the left ventricle, and the aorta The concentration of the dye in the aorta seldom becomes great and the dye disappears from the aorta while it still lingers in the right side of the heart

DIAGNOSIS

Accurate diagnosis may be difficult. Cyanosis may or may not be present, it frequently becomes manifest when the patient is between two and five years of age Fatigue, not dyspnea, limits the child s activity The great right sided car

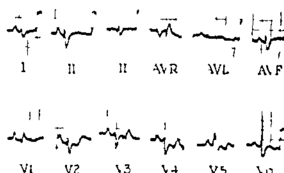


FIGURE XIX–7 Ebstein's anomaly of the tricuspid valve (standardized at 1 cm 1 millivolt) (same patient as in Figure XIX–2) Child

same may be true in V_2 and occasionally even in V_3 The deflections over the left precordium are, however, normal Figure XIX–7 shows the characteristic electrocardiographic changes When present, such findings offer strong confirmatory evidence It is, however, worthy of note that the author knows of two young infants with Ebstein's anomaly whose electrocardiograms were entirely normal

In one instance the displacement of the tricuspid valve was so extreme that the infant lived but three days, nevertheless, the unipolar precordial leads all showed entirely normal voltage

Yater and Shapiro[2] have reported a case in which from time to time there were changes in the shape of the ventricular complexes They believed that these changes were due to the variability in the course of the excitation wave associated with the anomalous structure of the right ventricle

SPECIAL TESTS

The circulation time is usually prolonged Because of the difficulty in the expulsion of blood from the right auricle and the right ventricle, the circulation time may be exceptionally long Even if the foramen ovale is patent, only rarely is a sufficient amount of test material shunted into the left auricle and thence into the systemic circulation to give a short circulation time

The oxygen saturation of the arterial blood varies with the volume of the right to-left shunt If the foramen ovale is sealed, the oxygen saturation of the arterial blood is normal If the foramen ovale is widely patent, a considerable amount of reduced hemoglobin will be shunted into the systemic circulation and there will be a proportional reduction in the oxygen saturation of the arterial blood

Cardiac catheterization is of aid when both the right auricle and the right ventricle are catheterized The pressure in the right auricle is usually increased,

diac enlargement, the poor quality of the heart sounds, the blurred and confused murmurs, the presence of a to-and fro murmur, and the occurrence of severe cardiac failure with engorgement of the liver but without pulsations at its margin suggest the diagnosis If the electrocardiogram shows low voltage in V_1 and normal deflections in V_5 and V_6, the diagnosis can be made with relative certainty The diagnosis is substantiated by the finding of a long circulation time and by angiocardiographic evidence of a huge right auricle in which the dye lingers for a long period Cardiac catheterization, if successful, shows an elevated pressure in the right auricle and a normal pressure in the right ventricle and the intracardiac electrocardiogram shows a ventricular type curve in the lower part of the auricle

DIFFERENTIAL DIAGNOSIS

In the absence of cyanosis Ebstein s anomaly must be differentiated from acute rheumatic fever, pericarditis, Lutembacher s syndrome, Fiedler s myocarditis, and at times from persistent patency of the ductus arteriosus In the presence of cyanosis it closely resembles isolated valvular pulmonary stenosis and may require differentiation from a tetralogy of Fallot.

Acute rheumatic fever may be confused with Ebstein's anomaly when there is a systolic murmur at the apex transmitted to the axilla and also a to-and fro murmur over the heart. The absence of symptoms of acute rheumatic fever such as fever, joint pains, rash, or abdominal pain and the finding of a normal sedimentation rate, as well as the clinical course, help to differentiate the two conditions

Pericarditis and pericardial effusion may be considered because of the to-and fro murmur and the enormous size of the heart. The long history of easy fatigability, the absence of other signs or symptoms of rheumatic fever or tuberculosis, and the knowledge that the condition dates from infancy or early childhood each offers a clue to the correct diagnosis

Lutembacher s syndrome that is, an auricular septal defect combined with mitral stenosis may also be considered Patency of the foramen ovale in Ebstein's anomaly causes a reduction in the pulmonary blood flow The tremendous dilatation of the pulmonary artery and the conspicuous hilar dance so characteristic of Lutembacher s syndrome never occur in Ebstein's anomaly

Fiedler s myocarditis may be difficult to differentiate from Ebstein's anomaly In Fiedler s myocarditis murmurs are absent and a pericardial friction rub is rare In the chronic form of interstitial myocarditis, exacerbations and remissions are common



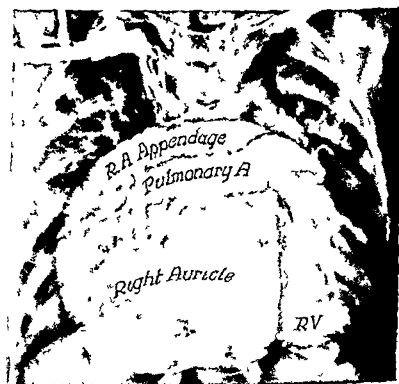


FIGURE XIX-8 Suspected Ebstein s anomaly of the tricuspid valve Child

PROGNOSIS

The prognosis is guarded The greater the distortion of the tricuspid valve and the larger the portion of the right ventricle which *lies above the valve*, the more serious is the prognosis The condition leads to progressive cardiac enlargement and ultimately to cardiac failure Furthermore, *sudden death* [8] is [illegible] due to ventricular fibrillation, to ventricular [illegible]

[illegible]

The cause is not known Nevertheless, the fear of sudden death [illegible] allowed to cast a gloom over the patient's life Many of us die suddenly, especially from automobile accidents The sudden death in a patient with Ebstein's anomaly is the mode of death for which many people long and with which few are blessed The author cared for one child who died so quietly, while talking happily with the other children in a pediatric ward, that when she stopped talking they thought she was "playing possum," and none of them knew that she had died The other children were merely told that she had gone to sleep and that in order to let her sleep she was carried out of the room

SUMMARY

In Ebstein's anomaly the tricuspid valve is malformed One or more leaflets of the tricuspid valve become fused with the endocardium of the right ventricle and a new valve is formed deep within that chamber The new valve divides the right ventricle into two chambers the upper part becomes incorporated into the right auricle and the lower part remains as the functional portion of the right ventricle The wall of the auricularized portion of the right ventricle is abnormally thin, whereas that of the outflow tract of the right ventricle is of approximately normal thickness Although the malformation seriously impairs the efficiency of both the right auricle and the right ventricle, the course of the circulation is normal except when the foramen ovale remains patent. Under such circumstances the increased pressure in the right auricle forces the valve covering the foramen ovale to open and a right to-left shunt is established

Fatigue is more marked than dyspnea Palpitation is a common complaint. Cardiac arrhythmias of all types are common

The presence or absence of cyanosis depends upon the patency of the foramen ovale and the volume of reduced hemoglobin shunted through it into the systemic circulation The greater the patency of the foramen ovale, the earlier is the appearance of cyanosis Cyanosis usually becomes manifest between two and five years of age The intensity of the cyanosis varies inversely with the volume

Persistent patency of the ductus arteriosus should not be confused with Ebstein's anomaly It is only in the presence of a to-and fro murmur that such a diagnosis may be entertained Certainly a definitive diagnosis of a patent ductus arteriosus should not be made in the presence of an acute pericarditis

Valvular pulmonary stenosis with an intact ventricular septum and the foramen ovale held open by the high pressure in the right auricle may closely simulate Ebstein's anomaly with a similar patency of the foramen ovale In Ebstein's anomaly the heart sounds are weak and confused, whereas in pulmonary stenosis the heart sounds are usually strong In both malformations the liver may be greatly enlarged but pulsations at the margin of the liver are the exception in Ebstein's anomaly and the rule in valvular pulmonary stenosis Angiocardiography usually indicates the thickness of the right ventricle in isolated valvular pulmonary stenosis the right ventricle is hypertrophied, whereas in Ebstein's anomaly it is exceptionally thin Cardiac catheterization in isolated valvular pulmonary stenosis reveals high pressure in both the right auricle and the right ventricle, whereas in Ebstein's anomaly the right auricular pressure is raised but the right ventricular pressure is normal

Tetralogy of Fallot may be confused with Ebstein's anomaly only if the new valve is formed deep within the right ventricle and the foramen ovale is functionally patent Under such circumstances the patient shows persistent cyanosis Moreover, when there is great distortion of the tricuspid valve, difficulty is encountered in the propulsion of blood into the right ventricle and, consequently, into the pulmonary artery Indeed, the abnormal tricuspid valve may obstruct the flow of blood into the outflow tract of the right ventricle and thereby cause functional pulmonary stenosis The great cardiac enlargement offers a clue to the correct diagnosis The engorgement of the liver and the electrocardiographic evidence of intraventricular conduction disturbance immediately suggest that the malformation is not a tetralogy of Fallot The determination of the circulation time may be of real value in the differentiation of a tetralogy of Fallot from Ebstein's anomaly In a tetralogy of Fallot the circulation time is abnormally short and in Ebstein's anomaly it is usually abnormally long When the circulation time is approximately normal, angiocardiography may be necessary to determine the presence or absence of an over riding aorta

TREATMENT

There is none Digitalis may help to maintain compensation and to control the heart rate and thereby to lessen the incapacity due to cardiac arrhythmias Cardiac failure should be treated in the usual manner

PROGNOSIS

The prognosis is guarded The greater the distortion of the tricuspid valve and the larger the portion of the right ventricle which *lies above* the valve, the more serious is the prognosis The condition leads to progressive cardiac enlargement and ultimately to cardiac failure Furthermore, *sudden death*[3] is relatively common It may be due to ventricular fibrillation, to ventricular tachycardia, to prolonged auricular tachycardia, or even to ventricular standstill The cause is not known Nevertheless, the fear of sudden death should never be allowed to cast a gloom over the patient s life Many of us die suddenly, especially from automobile accidents The sudden death in a patient with Ebstein s anomaly is the mode of death for which many people long and with which few are blessed The author cared for one child who died so quietly, while talking happily with the other children in a pediatric ward, that when she stopped talking they thought she was playing possum, and none of them knew that she had died The other children were merely told that she had gone to sleep and that in order to let her sleep she was carried out of the room

SUMMARY

In Ebstein s anomaly the tricuspid valve is malformed One or more leaflets of the tricuspid valve become fused with the endocardium of the right ventricle and a new valve is formed deep within that chamber The new valve divides the right ventricle into two chambers the upper part becomes incorporated into the right auricle and the lower part remains as the functional portion of the right ventricle The wall of the auricularized portion of the right ventricle is abnormally thin whereas that of the outflow tract of the right ventricle is of approximately normal thickness Although the malformation seriously impairs the efficiency of both the right auricle and the right ventricle, the course of the circulation is normal except when the foramen ovale remains patent Under such circumstances the increased pressure in the right auricle forces the valve covering the foramen ovale to open and a right to-left shunt is established

Fatigue is more marked than dyspnea Palpitation is a common complaint Cardiac arrhythmias of all types are common

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of the right-to-left shunt Cyanosis is usually pronounced when the foramen *ovale is widely patent*

The greater the displacement of the tricuspid valve, the greater is the enlargement of the heart The heart sounds are weak, the murmurs are blurred and confused There is often a to-and-fro murmur which simulates a pericardial friction rub The liver becomes engorged and there may be peripheral edema

The electrocardiogram is of a great diagnostic aid, the deflections in V_1 are of low amplitude and of long duration and those in V_5 and V_6 are of normal height

The circulation time is markedly prolonged

Angiocardiography reveals a huge right auricle and a thin walled right ventricle The dye lingers for a long time in the right side of the heart

Cardiac catheterization shows a relatively high pressure in the right auricle and a normal pressure in the right ventricle, an intracardiac electrocardiogram shows deflections which are characteristic of those obtained from the ventricle while the catheter is still in the auricle

The malformation must be differentiated from acute rheumatic fever, pericarditis, Lutembacher's syndrome, persistent patency of the ductus arteriosus, isolated valvular pulmonary stenosis, and a tetralogy of Fallot

Accurate diagnosis is important, as this malformation is not amenable to surgery Most patients live to adolescence or to adult life A peaceful sudden death is relatively common

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CHAPTER XX

PERSISTENT PATENCY OF THE DUCTUS ARTERIOSUS

THE persistence of the ductus arteriosus in adult life produces a clinical syndrome which in its most characteristic form is extremely distinctive and hence one of the easiest to diagnose with certainty

The etiology is obscure The evidence is, however, clear that infection of the mother with a rubella virus during the first trimester of pregnancy frequently injures the fetus The resultant defects are usually microcephaly, congenital cataracts, and persistent patency of the ductus arteriosus (see Chapters I and V)

NATURE OF THE MALFORMATION

The ductus arteriosus is a normal fetal pathway It is given off from the pulmonary artery at the bifurcation of the two main branches and opens into the aorta beyond the origin of the left subclavian artery The failure of the ductus arteriosus to undergo obliteration results in the persistence of a vessel which connects the aorta with the pulmonary artery, as shown in Figure XX–I

During fetal life the lungs do not function and the pressure in the pulmonary vascular bed is relatively high The ductus arteriosus serves to direct the greater part of the blood which is pumped into the pulmonary artery to the systemic circulation

At birth the ductus arteriosus is normally patent Shortly after birth it begins to undergo obliteration The process is usually complete by the end of the second month but occasionally the closure of the ductus arteriosus may be delayed for six months or even for a year Therefore, if the ductus is normal in size and position and the remainder of the cardiovascular system is normal, simple patency of the ductus arteriosus during the first months of life should not be considered an abnormality but rather a normal variant

Functional closure of the ductus arteriosus usually occurs at an early age, exactly how early is not known It is generally assumed that the murmur which occurs during the neonatal period and subsequently disappears is due to the flow of blood through the ductus arteriosus It is probable that the disappearance of the murmur coincides with the functional closure of this pathway Such a murmur is not heard in every heart It may well be that the absence of a murmur

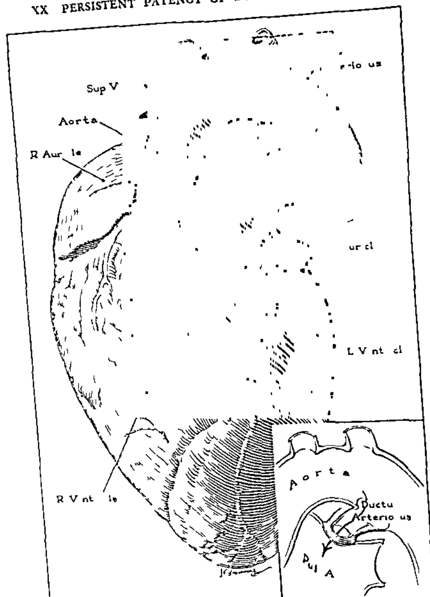


FIGURE XX-1 Patent ductus arteriosus Adult

means that there is no significant flow of blood through the ductus arteriosus The work of Barclay et al[1 2] indicates that, at least in the sheep s heart, functional closure of the ductus arteriosus usually occurs within the first few cycles of extra uterine life

On the other hand the catheterization studies by Adams and Lind[3] indicate

that even in the absence of a murmur a considerable volume of blood may flow from the aorta through the ductus arteriosus for several days

Although the time at which the flow of blood through the ductus arteriosus normally ceases may be subject to some variation, the physiological patency in early infancy is to be clearly differentiated from the persistent patency of the ductus arteriosus throughout life Furthermore, when the ductus arteriosus does remain patent, it is important to appreciate that in some instances it adds to the strain on the circulation and in other instances, as in a tetralogy of Fallot with pulmonary atresia, it constitutes the main pathway by which the blood reaches the lungs

The present discussion is concerned primarily with persistent patency of the ductus arteriosus when it occurs as an isolated abnormality

The inherent tendency of the ductus arteriosus to obliterate appears to be a fundamental characteristic of this tissue Kennedy[4] and Kennedy and Clark[5, 6] have presented experimental evidence which indicates that the oxygen content of the blood is an important factor in the obliteration of the ductus arteriosus Certain it is that persistence of the ductus arteriosus does not depend primarily upon its size, on the relative pressure in the two circulations, or on the physiological importance of the pathway There are a number of malformations of the heart in which the ductus arteriosus is the only pathway to the lungs and in which, even though the closure of the ductus may be delayed, it eventually undergoes obliteration, thereby rendering the condition incompatible with life On the other hand, there are some instances in which the ductus arteriosus, although unimportant to the maintenance of life, fails to undergo normal obliteration In these instances it seems as though the tissue lacked the inherent tendency to undergo obliteration A deficiency of such an inherent tendency in the tissue would explain the frequency with which the persistent patency of the ductus arteriosus occurs as an isolated malformation Inasmuch as the obliteration of the ductus arteriosus is a normal phenomenon, when the ductus remains patent after two years of age, it represents an anomaly whether or not it counterbalances the effect of another malformation or occurs as an isolated abnormality

Usually the obliteration of the ductus arteriosus starts at the pulmonary end and eventually extends throughout the length of the ductus Sometimes the ductus arteriosus remains as a strand of tissue but it often atrophies and completely disappears Occasionally the closure of the ductus arteriosus is accomplished by the formation of a membrane across the pulmonary end of the opening Twice at autopsy the author has seen such a membrane, which did not entirely close

the opening It is conceivable that with the rise in the systemic pressure the membrane could rupture Such a phenomenon would account for the occasional abrupt appearance of the signs of patency of the ductus arteriosus in late childhood or early adult life

COURSE OF THE CIRCULATION

During fetal life the ductus arteriosus permits the blood to flow from the pulmonary artery to the descending aorta Inasmuch as this vessel is a normal fetal pathway, it places no abnormal strain upon the fetal heart

At birth, with the expansion of the lungs, the pulmonary pressure falls and the blood in the pulmonary artery is directed to the lungs, an increased volume of blood is returned from the lungs to the left auricle The pressure in the left auricle rises and the valve which covers the foramen ovale floats to closure Very shortly thereafter the pulmonary pressure falls, the pressure in the systemic circulation rises and becomes relatively greater than the pressure in the lesser circulation and the direction of the flow of blood through the ductus is *reversed* that is, the blood instead of flowing as it does during fetal life from the pulmonary artery to the aorta, now flows from the aorta to the pulmonary artery

In the normal heart all experimental evidence[1, 3] indicates that the pressure in the two circulations subsequently becomes equal, regardless of whether or not blood flows through the ductus arteriosus Thereafter the ductus arteriosus undergoes obliteration Thus functional closure of the ductus arteriosus precedes the anatomical closure of this pathway

When the ductus arteriosus fails to undergo normal obliteration, as the pressure in the systemic circulation becomes higher than that in the pulmonary circulation, blood again flows from the aorta through the ductus to the pulmonary artery The volume of blood so shunted to the lungs will vary directly with the size of the ductus If the ductus is abnormally large, the pulmonary blood flow will be proportionally increased All present evidence indicates that under such circumstances the pulmonary vascular bed does not expand as rapidly as it does in the normal infant[7] Consequently the pulmonary resistance remains abnormally high This increased resistance in the pulmonary vascular bed is a protective mechanism to prevent an excessively, possibly even a fatally, large blood flow to the lungs In general the larger the ductus, the more gradual is the expansion of the pulmonary vascular bed and the longer the pulmonary hypertension persists

It is, however important to remember that in the presence of a large ductus

it is the large arteriovenous shunt which causes the pulmonary hypertension and that this, in turn, permits an equilibrium to be established between the two circulations Consequently, even though the pressures in the two circulations are approximately equal, the predominant direction of the shunt is from the aorta to the pulmonary artery

The course of the circulation is shown in Diagram xx–1 The blood from the right auricle passes into the right ventricle and is pumped out by way of the pulmonary artery to the lungs The oxygenated blood from the lungs is returned by the pulmonary veins to the left auricle Thence it flows to the left ventricle and is pumped out by way of the aorta to the systemic circulation Inasmuch as the pressure is normally higher in the aorta than in the pulmonary artery, some blood flows through the ductus arteriosus to the pulmonary artery This blood is again pumped around the lesser circulation and is returned to the left auricle and the left ventricle, thence it is again pumped out into the aorta

PHYSIOLOGY OF THE MALFORMATION

Persistent patency of the ductus arteriosus causes a left to right shunt and increases the work of both ventricles The blood which is shunted through the ductus arteriosus to the lungs increases the volume of blood which is returned to the left auricle and the left ventricle These chambers become enlarged The increased volume of blood in the pulmonary circulation also slightly increases the work of the right ventricle, the right ventricle, too, becomes slightly enlarged Consequently there is dilatation and hypertrophy of the left auricle and of both ventricles

The greater the volume of the shunt, the greater is the load placed upon the heart and the pulmonary circulation and the more gradually does the pulmonary vascular bed open Nevertheless, the pulmonary vascular bed usually opens up in the normal manner and even a large ductus, when it occurs as an isolated malformation, seldom causes persistent pulmonary hypertension

SEX INCIDENCE

In this malformation, as in a number of others, there is a curious sex distribution For some unknown reason the malformation is two or three times more common in females than in males [8]

CLINICAL FINDINGS

The symptoms produced by a patent ductus arteriosus vary with the size of the ductus When the ductus arteriosus is small, the patient may be entirely

asymptomatic When the ductus arteriosus is large, there may be a considerable degree of incapacity

Dyspnea and *palpitation* are the outstanding symptoms in patients with great cardiac enlargement Infants with a huge patent ductus arteriosus may suffer from polypnea and repeated respiratory *infections*

Failure to gain and *stunting of growth* occasionally occur when there is an enormous flow of blood through the ductus arteriosus to the lungs and a proportional decrease in the blood supply to the systemic circulation

Fortunately in the majority of cases the ductus arteriosus is relatively small and the symptoms produced thereby are minimal Frequently the condition is first detected in the course of a routine physical examination

Cyanosis seldom occurs in this malformation The direction of the shunt is always from the area of high pressure to that of low pressure, hence the flow of blood is from the systemic to the pulmonary circulation Arterial blood is shunted into the pulmonary artery but venous blood is *not ordinarily shunted* into the aorta, hence there is no cyanosis It is only when the pressure in the lesser circulation is abnormally high, as may occur in infants with a large ductus arteriosus, that some venous blood may be shunted into the systemic circulation [9] Under such circumstances an infant may show slight cyanosis Such cyanosis usually decreases with crying

In adults with a normal pulmonary vascular bed a reversal in the direction of the shunt virtually never occurs

Clubbing of the extremities never occurs in a patient with an uncomplicated patency of the ductus arteriosus

Aphonia and *hoarseness* are rare complaints but may occur when an enormously dilated pulmonary artery presses on the recurrent laryngeal nerve

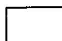
The pulse has a quick rise and a quick fall, which makes it feel like a wire *whipping* against one s finger This is caused by the unusually *wide pulse pressure* The normal pulse pressure is less than 40 mm of mercury, when the ductus arteriosus remains patent, the pulse pressure increases in width The diastolic pressure is usually low and upon exercise may drop to zero The pulse pressure may be so wide and the diastolic pressure so low as to produce peripheral signs of aortic insufficiency with a pulse of the Corrigan type and a conspicuous capillary pulse in the nail beds

CARDIAC FINDINGS

A continuous machinery murmur is the most characteristic of all findings in a patent ductus arteriosus It denotes the continuous flow of blood from one

Diagram xx–1

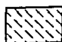
Head and upper extremities
Ductus Arteriosus
R Lung
P.A.
S.V.C.
Aorta
Pul A
L Lung
PV
Pul V
Pulmonary Art
L A
R A
L V
R.V.
Descending Aorta
I.V.C.
Trunk and lower extremities



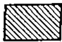
Arterial blood (fully saturated)



Venous and arterial blood
Cyanosis visible



Small admixture of venous blood
No visible cyanosis



Venous blood

DIAGRAM XX-1

Persistent patency of the ductus arteriosus

In this anomaly the ductus arteriosus remains patent throughout life

The blood from the right auricle passes to the right ventricle and out by way of the pulmonary artery to the lungs The oxygenated blood from the lungs is returned in the normal fashion by the pulmonary veins to the left auricle, thence it passes to the left ventricle and is pumped out through the aorta to the systemic circulation Inasmuch as the pressure in the aorta is greater than that in the pulmonary artery, the blood flows from the aorta through the ductus arteriosus to the pulmonary artery The blood so shunted flows out through the pulmonary arteries to the lungs and is returned to the left auricle, thence it flows to the left ventricle and is again pumped out into the aorta The increased volume of blood in the aorta raises the pressure in that vessel, blood is again shunted through the ductus arteriosus into the pulmonary artery Thus a shunt is established through the lesser circulation and the work of the left auricle and the left ventricle is increased The increased pressure in the pulmonary artery raises the pressure in the pulmonary circulation and causes the right ventricle to work against an abnormally high pressure The right auricle is spared Hence it is common to find dilatation of the pulmonary artery proximal to the ductus arteriosus, usually the right ventricle is slightly hypertrophied

Clinical diagnosis The characteristic findings on physical examination are a continuous murmur and a thrill over the pulmonary area The murmur is best heard in the second left interspace above the area of cardiac dullness The murmur usually has a systolic accentuation but extends throughout diastole, the systolic element of the continuous murmur is transmitted to the vessels of the neck and is audible posteriorly, high in the interscapular region The size of the heart is determined by the volume of the shunt Both ventricles have an increased amount of work and there is usually slight symmetrical enlargement of both sides of the heart. Inasmuch as the shunt is from the aorta to the pulmonary artery, there is no cyanosis and no clubbing The electrocardiogram usually shows a balanced axis and evidence of combined hypertrophy or a left ventricular dominance

circulation to the other The murmur is of a harsh, rasping quality and is continuous throughout both systole and diastole Usually the murmur has a systolic accentuation Such a murmur is commonly described as a 'machinery' murmur Although some observers consider any murmur which is extremely harsh and rasping a "machinery" murmur, the term is generally used to describe the murmur characteristic of a patent ductus arteriosus Therefore this term should be limited to a *continuous* murmur, that is, a murmur which resembles a machine running continuously When so used, the term "machinery" murmur is always suggestive of persistent patency of the ductus arteriosus

The development of the 'machinery murmur depends upon the relative pressure in the two circulations Inasmuch as in infancy the systolic pressure is low and the pulmonary pressure relatively high, the difference in the pressure between the two circulations is slight, consequently during this age period there is usually only a systolic murmur With the growth of the child the pressure in the systemic circulation rises and the pressure in the lesser circulation gradually falls When the diastolic pressure in the systemic circulation becomes greater than the diastolic pressure in the pulmonary circulation, then and then only does the patient develop the continuous machinery' murmur so characteristic of a patent ductus arteriosus Occasionally this change in pressure occurs abruptly after an intercurrent illness Under such circumstances the continuous murmur develops with corresponding abruptness If, however, the pulmonary pressure is abnormally high, the development of the continuous murmur will not occur until the systemic pressure has risen to a proportionally higher level

The intensity of the murmur is directly proportional to the volume of the shunt and this, in turn, depends upon the size of the ductus and on the difference in pressure between the systemic and the pulmonary circulation Thus, in the absence of pulmonary hypertension, the intensity of the murmur and the size of the heart varies with the size of the ductus When the ductus is small, the heart is normal in size and the continuous murmur is relatively faint When, however, the ductus is abnormally large, the heart is greatly enlarged and the continuous murmur is proportionally intense It is important to remember that, when the ductus is extremely large, the child may be four, five, or even six years of age before the development of the characteristic continuous murmur During the intervening period a precordial systolic murmur is the rule and frequently there may be a prolonged *mid diastolic murmur* and a *gallop rhythm* due to the poorly functioning heart

The absence of the characteristic machinery murmur in infancy *and in*

early childhood renders the diagnosis of a patent ductus arteriosus extremely difficult at this age Fortunately most infants with persistent patency of the duc tus arteriosus do well and early diagnosis is not necessary *Occasionally*, however, a large ductus may cause great incapacity Under such circumstances special tests are indicated (see below)

The following sequence of events is relatively common A doctor who has observed an infant from birth may note toward the end of the first year the de velopment of a systolic murmur At first the murmur is indistinguishable from a functional murmur, later it becomes so loud as to arouse suspicion of a con genital malformation of the heart Nevertheless, several years may elapse before the murmur develops the machinery quality which is so characteristic of a patent ductus arteriosus Although until the development of the continuous murmur a definitive diagnosis cannot be made without special *studies*, there after the condition becomes one of the simplest to diagnose clinically

The location and the transmission of the murmur as well as its quality are quite characteristic The continuous murmur is best heard in the second left in terspace beneath the clavicle The murmur is normally maximal to the left of the sternum because the ductus arteriosus extends from the pulmonary artery to the descending aorta, which normally lies to the left of the spinal column The author was told of one patient[10] with a right aortic arch in whom the ductus ar teriosus persisted on the right side instead of on the left side In this instance, in which the patency of the ductus arteriosus and the right aortic arch were the only abnormalities, the murmur and thrill were maximal to the right of the sternum

The systolic element of the murmur is ordinarily transmitted to the vessels of the neck and is usually clearly audible posteriorly, high in the interscapular region Almost invariably the systolic murmur is audible over the entire pre cordium Indeed, the systolic murmur may be loudest in the third left interspace close to the sternum and may even be transmitted to the axilla Moreover, the *diastolic component* can often be heard along the left sternal border Careful examination will always show that there is a *continuous* murmur of maximal intensity to the left of the sternum beneath the clavicle This murmur is usually louder in the recumbent than in the erect position and is increased in intensity by exercise

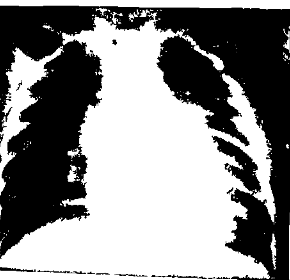
A thrill as well as a murmur, is usually present It, too, may be continuous and of maximal intensity over the pulmonary area, that is, high in the second left interspace beneath the clavicles Indeed, the location and the quality of the thrill are so characteristic that frequently the diagnosis can be made by palpation

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The absence of the characteristic machinery murmur in infancy and in



Anterior posterior position



Right anterior-oblique position

Figure xx-3 Patent ductus arteriosus Infant

ductus is nearly as large as the descending aorta Dammann and his associates[11] were the first to report a series of infants who suffered from tremendous pulmonary blood flow Under such circumstances the pulmonary pressure is high, there is no continuous murmur, but a systolic and a mid diastolic murmur are present over the precordium and frequently a gallop rhythm is audible at the apex Special tests are usually needed to establish the diagnosis

In rare instances cardiac failure may occur in childhood as the pulmonary vascular bed expands and the volume of the shunt is thereby increased When the heart is greatly enlarged, any superimposed infection which increases the work of the heart may be sufficient to cause cardiac failure

Cardiac failure also occurs in adults with a large ductus arteriosus in whom the heart is greatly enlarged Such a patient has a low cardiac reserve The wide pulse pressure with a low diastolic pressure reduces the efficiency of the coronary circulation These factors, together with the stress and strain of advancing years, may lead to cardiac failure, with or without auricular fibrillation Now that the condition can be corrected by surgery, this late complication is preventable

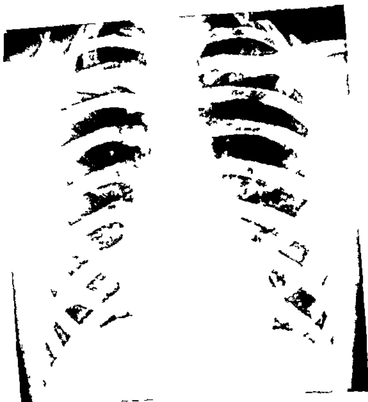
Auricular fibrillation is a rare late complication, but may occur in an adult whose heart is greatly enlarged This difficulty, too, should be obviated by early surgery

X RAY AND FLUOROSCOPIC FINDINGS

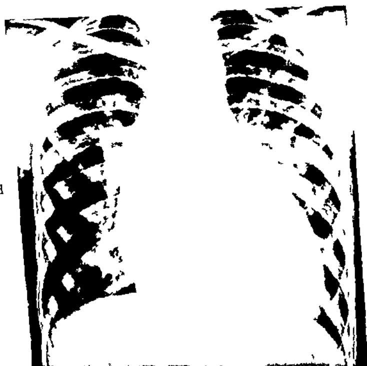
The contour of the heart is subject to considerable variation The feature most commonly described as characteristic of the malformation is the accentuation of the second curve to the left of the sternum This shadow may be caused by the dilatation of the pulmonary artery proximal to the ductus arteriosus Generally it is due to the presence of a large thymus This curve, known as the "cap of Zinn," is the rule in infants and young children and is rarely seen in older individuals The disappearance of this shadow is undoubtedly due to the fact that at puberty the thymus gland decreases in size The characteristic contours of the heart in infants, children, and adults are shown in Figures XX–3 through 5

These x rays illustrate that, although prominence of the pulmonary conus is common with persistent patency of the ductus arteriosus, it does not always occur The converse is also true, there may be prominence of the pulmonary conus with no underlying anomaly Indeed, slight prominence of the pulmonary conus is common in children Therefore the diagnosis of a patent ductus arteriosus should never be made solely on the basis of the contour of the heart

The hilar vascularity is generally increased and the hilar shadows are more

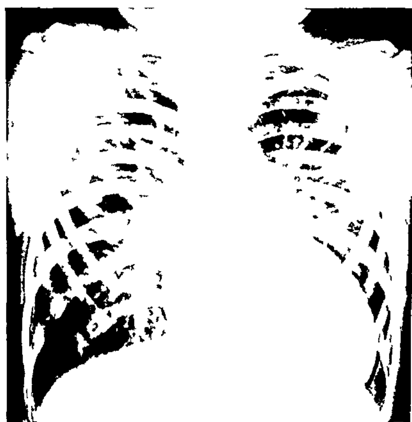


Adult



Child

FIGURE XX–5 Patent ductus arteriosus without prominence of the pulmonary conus



Child



Child

FIGURE XX-4 Patent ductus arteriosus showing a prominent cap of Zinn Two cases

is evidence of left ventricular strain In childhood slight prolongation of the P R interval is common but the electrocardiogram is usually normal, although the precordial leads may show evidence of slight left ventricular dominance

SPECIAL TESTS

The red blood cell count, the level of the available hemoglobin, and the hematocrit reading are within normal limits The oxygen saturation of the arterial blood is usually normal Occasionally in infants with a large heart and marked pulmonary hypertension there may be slight oxygen unsaturation of the arterial blood,[11] when such unsaturation is present, it is usually lessened by exercise

Cardiac catheterization may be necessary to establish the diagnosis before the development of a continuous murmur The oxygen content of the blood in the right auricle should be the same as that in the superior vena cava, that in the right ventricle may be higher than that in the right auricle The greatest increase in the oxygen content of the blood should occur in the sample taken from the pulmonary artery Prior to the development of a continuous murmur the pulmonary pressure is usually elevated It may or may not be possible to pass the catheter through the ductus arteriosus into the descending aorta In general the higher the pulmonary pressure, the easier it is to catheterize the ductus If the catheter is passed below the diaphragm, such a procedure is proof positive of the existence of a ductus but it does not prove that patency of the ductus arteriosus is the only abnormality

Angiocardiography is of greater aid in the exclusion of other malformations than in the diagnosis of a simple patency of the ductus arteriosus

Aortography is probably the most useful diagnostic procedure for the establishment of the diagnosis of patency of the ductus arteriosus in infants In order to demonstrate passage of the dye from the aorta to the lungs the films must be taken in rapid succession, and the first film must be taken as the dye is injected The dye may be so rapidly dissipated that the films may require careful study in order to see the dye in the lungs If there is no communication between the aorta and the pulmonary artery, no dye will enter the lesser circulation The presence of dye in the lungs, as shown in Figure xx–7, offers positive proof of a direct communication between the aorta and the pulmonary artery It does not, however, exclude the possibility of an aortic septal defect or of a truncus arteriosus, nor does it prove that patency of the ductus arteriosus is the sole abnormality

conspicuous Inasmuch as the volume of the pulmonary blood flow varies with the size of the ductus arteriosus, the larger the ductus, the greater is the cardiac enlargement

Upon fluoroscopy the shape of the heart may be essentially normal but both ventricles may be slightly enlarged The pulmonary conus may or may not be prominent Slight enlargement of the left auricle is common, especially in infants and young children, as the diaphragm lies at a high level, consequently left auricular enlargement is more easily detected at this age (see Figure xx-3) Usually there is a conspicuous pulsation in the region of the pulmonary artery and the hilar vascularity is increased Although a patent ductus arteriosus is one of the conditions which cause a "hilar dance," it is important to remember that the most conspicuous pulsations in the pulmonary conus and in the hilar shadows occur, not in a patent ductus arteriosus, but in anomalies of the venous return (see Chapter xxxii)

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram varies with the age of the patient and the size of the ductus arteriosus In infants with a large ductus arteriosus, the unipolar precordial leads show evidence of "combined" hypertrophy, that is, evidence of right ventricular hypertrophy in V_1 and left ventricular dominance in V_6, as shown in Figure xx-6A In some instances there may be a left axis deviation and evidence of left ventricular hypertrophy (see Figure xx-6B) Occasionally there

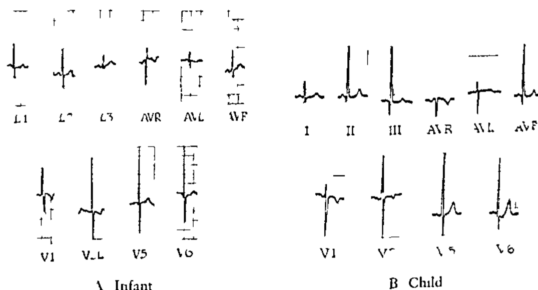


Figure xx-6 Patent ductus arteriosus

DIAGNOSIS

Diagnosis may be suspected when a child with cataracts *has a cardiac abnormality* and the mother gives a history of rubella in early pregnancy

Prior to the development of a continuous murmur the condition cannot be clinically diagnosed with certainty It is, however, to be suspected in an infant who has a wide pulse pressure, a large heart with a systolic and a *diastolic* murmur over the precordium, and a gallop rhythm The diagnosis is *further* subs antiated when fluoroscopy shows enlargement of the left auricle and both ventricles and increased vascular markings and when the electrocardiogram shows a balanced axis in the standard leads and "combined' hypertrophy in the unipolar precordial leads

Infants in whom a large ductus arteriosus causes difficulty in early life frequently have some additional malformation If, however, the two malformations are additive, removal of the load placed on the heart by the ductus arteriosus may enable the infant to adjust to the other malformation Therefore, if the existence of a large patent ductus arteriosus is suspected, and especially if there is evidence of chronic cardiac failure, additional studies *are strongly indicated*

After the development of a continuous murmur the clinical diagnosis can be made on the basis of a continuous murmur over the pulmonary area in a patient who shows no cyanosis and has a balanced electrocardiogram with evidence of combined' hypertrophy in the precordial leads The *shunt is always from the* aorta to the pulmonary artery In cases in which the volume of the *shunt is large, the diastolic* pressure is low and the pulse pressure wide Such patients may have considerable cardiac enlargement and show peripheral signs of aortic insufficiency Fluoroscopy may reveal pulsations of the hilar shadows

When the diagnosis is based on these simple clinical findings, the errors in diagno is are *less than 2 per cent* If all known laboratory tests are used, these errors can be reduced by only 1 per cent The risks and discomforts of these *tests* are sufficiently great so that extensive investigation is not warranted merely *to* improve accuracy in diagnosis

DIFFERENTIAL DIAGNOSIS

Although in its most characteristic form persistent patency *of the* ductus arteriosus is relatively easy *to* diagnose, there are several conditions *from* which it must be differentiated

Prior to the development of a continuous *murmur the condition* requires differentiation from other malformations *which cause* a large left to-right shunt

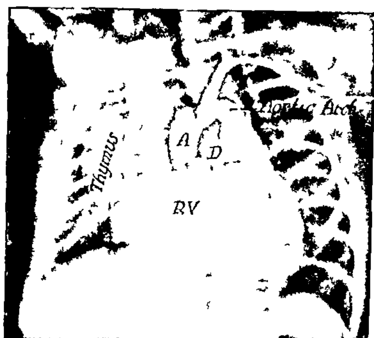




FIGURE XX–7 Patent ductus arteriosus Infant

DIAGNOSIS

Diagnosis may be suspected when a child with cataracts has a cardiac abnormality and the mother gives a history of rubella in early pregnancy

Prior to the development of a continuous murmur the condition cannot be clinically diagnosed with certainty It is, however, to be *suspected in an infant* who has a wide pulse pressure, a large heart with a systolic and a diastolic murmur over the precordium, and a gallop rhythm The diagnosis is further substantiated when fluoroscopy shows enlargement of the left auricle and both ventricles and increased vascular markings and when the electrocardiogram shows a balanced axis in the standard leads and 'combined hypertrophy in the unipolar precordial leads

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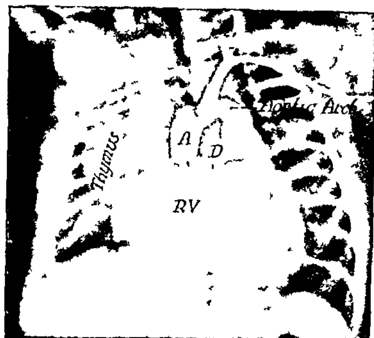
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When the diagnosis is based on these simple clinical findings, the errors in diagnosis are less than 2 per cent If all known laboratory tests are used, these errors can be reduced by only 1 per cent The risks and discomforts of these tests are sufficiently great so that extensive investigation is *not warranted* merely to improve accuracy in diagnosis

DIFFERENTIAL DIAGNOSIS

Although in its most characteristic form persistent patency of the ductus arteriosus is relatively easy to diagnose, there are several conditions from which it must be differentiated

Prior to the development of a continuous murmur the condition requires differentiation from other malformations which cause a large left to-right shunt



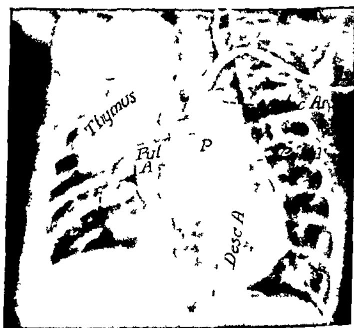


FIGURE XX–7 Patent ductus arteriosus Infant

ductus arteriosus is usually a relatively easy diagnosis There are, however, several conditions from which this malformation must be differentiated

A venous hum, which is of common occurrence in childhood, must not be confused with the continuous murmur of a patent ductus arteriosus A venous hum is a continuous murmur with a *diastolic* accentuation The murmur is frequently heard beneath the clavicle to the left of the sternum but is seldom re [illegible] also heard to the right of the sternum and [illegible]

heart, indeed it is often loudest directly over [illegible] hum may be extraordinarily loud It is, however, louder in the erect position than in the recumbent position This phenomenon is diametrically opposite to the changes in the intensity of the murmur found in a patent ductus arteriosus A venous hum may or may not be obliterated by pressure on the neck vessels The most characteristic of all features of a venous hum is its change in intensity with changes in the position of the head and neck

Severe anemia may cause murmurs which are confused with persistent patency of the ductus arteriosus Although these murmurs are usually systolic in time, in rare instances a diastolic murmur may be audible over the base of the heart Indeed, in the presence of a severe anemia, all murmurs must be evaluated with caution

An aortic septal defect may give exactly the same clinical and laboratory findings as simple patency of the ductus arteriosus Occasionally the murmur is maximal over the manubrium or to the right of the sternum In other instances the correct diagnosis cannot be made prior to operation If at operation no ductus is found, it is probable that the condition is due to an aortic septal defect Nevertheless exploration of the base of the aorta should be attempted only if the surgeon is prepared to close an aortic septal defect, and if the family is fully cognizant of the situation and of the risk of operation (see Chapter XXI)

A truncus arteriosus with adequate or excessive pulmonary blood flow may occasionally be confused with a patent ductus arteriosus It is the occurrence of cyanosis and of oxygen unsaturation of the arterial blood, combined with the wide transmission of the continuous murmur over the lungs, which gives the clue to the correct diagnosis

A hemi truncus arteriosus is a rare condition which may occasionally simulate a patent ductus arteriosus An uncomplicated hemi truncus means that the only abnormality in the cardiovascular system is that one pulmonary artery arises from the aorta and the other pulmonary artery arises in the normal man

namely, septal defects, both auricular and ventricular After the development of the continuous murmur the condition may be confused with a venous hum or with the hemic murmur associated with severe anemia The condition also requires differentiation from an aortic septal defect, a truncus arteriosus with adequate or excessive pulmonary blood flow, a hemi truncus arteriosus, primary pulmonary hypertension *with* or *without* patency of the ductus arteriosus, a high ventricular septal defect with aortic insufficiency, severe aortic insufficiency, rheumatic heart disease, *rupture of an aneurysm of the sinus of Valsalva into the* lesser circulation, and other rare conditions which cause a continuous murmur, and also from aortic or pulmonary insufficiency

An auricular septal defect may cause great cardiac enlargement and both a systolic murmur and a mid diastolic murmur at the apex An auricular septal defect is usually associated with electrocardiographic evidence of a right axis deviation and a right bundle branch block, whereas a patent ductus arteriosus is associated with evidence of a balanced axis and "combined' hypertrophy

A defect of the ostium primum type or a persistent ostium atrioventriculare commune may cause great cardiac enlargement and cardiac failure in early infancy An ostium primum defect causes a systolic murmur and a thrill which are maximal over the body of the heart and, although the electrocardiogram shows a left axis deviation, the precordial leads show evidence of an incomplete right bundle branch block A persistent ostium atrioventriculare commune may cause great difficulty in early infancy An aortogram may be necessary to differentiate these conditions from that of patency of the ductus arteriosus

A large ventricular septal defect with increased pulmonary blood flow and high pulmonary pressure more closely resembles patency of the ductus arteriosus than do either of the above malformations The electrocardiogram is of less diagnostic aid, as there may be either a left or a right axis deviation in the standard leads and evidence of preponderance of either left or right ventricular hypertrophy in the precordial leads Fluoroscopic examination will show cardiac enlargement, increased pulmonary blood flow, and upon barium swallow evidence of left auricular enlargement

These three malformations all may require special procedures to differentiate them from patency of the ductus arteriosus Cardiac catheterization will show whether the shunt occurs at the auricular or the ventricular level or not until the pulmonary artery is entered Aortography will show whether the shunt is above or below the aortic valve

After the development of the continuous murmur, persistent patency of the

pressure rises, it is usually *safe to* ligate the ductus If, however, the pressure in the pulmonary artery rises, closure of the *ductus arteriosus* should not be attempted

Furthermore, it should be remembered that there are certain persons in whom the two *conditions* exist together and in whom, although it may be possible to ligate the ductus successfully, over a period of years such a procedure may do more harm than good, because the escape valve for the pulmonary hypertension has been closed If a large *right to*-left shunt through the ductus arteriosus is present, closure of the ductus is usually strongly contraindicated

A high ventricular septal defect when combined with aortic insufficiency may be confused with persistent patency of the ductus arteriosus, as both conditions are associated with a large left to-right shunt, increased pulmonary blood flow, and both a systolic and a diastolic murmur In a high ventricular septal defect combined with *aortic insufficiency*, the quality of the murmur, as well as its location, is of diagnostic aid Usually the early diastolic murmur is of maximal intensity in the third and fourth left interspaces close to the sternum, whereas in a patent ductus arteriosus the murmur and the thrill are maximal higher up to the left of the sternum, and the murmur has a crescendo-decrescendo quality Cardiac catheterization or aortography is occasionally necessary to differentiate the two conditions

Aortic insufficiency when severe, occasionally may be confused with patency of the ductus arteriosus or with an aortic septal defect The occurrence of aortic insufficiency causes no shunt from left to right and therefore cardiac catheterization will reveal no abnormal findings but aortography will show reflux of blood into the left ventricle.

Rheumatic heart disease with both aortic and mitral insufficiency, is occasionally confused with patency of the ductus arteriosus Such errors are most likely to be made when the aortic diastolic murmur is maximal over the base of the heart and not along the left sternal border The systolic murmur is, however, always maximal at the apex and is well transmitted to the axilla In contrast to this, in a patent ductus arteriosus both the systolic and the diastolic elements of the murmur and the thrill are of maximal intensity at the base of the heart in the second left interspace beneath the clavicle Although the continuous murmur of a patent ductus arteriosus may mask the existence of aortic insufficiency, careful examination should enable the physician to distinguish between the two conditions

The rupture of an aneurysm of the sinus of Valsalva into the right auricle or right ventricle may produce a murmur similar to that of a patent ductus arterio-

ner from the right ventricle (see Chapter xiv) Generally the murmur of a hemi truncus arteriosus has a humming quality and is widely transmitted throughout the lung

Primary pulmonary hypertension which has progressed to such an extent as to cause pulmonary insufficiency may produce a systolic murmur and a diastolic murmur of such a nature as to simulate those of a patency of the ductus arteriosus The murmur is, however, usually maximal over the precordium rather than over the pulmonary area The pulmonic second sound is usually remarkably accentuated Furthermore, the electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy

Pulmonary hypertension combined with patency of the ductus arteriosus calls for differentiation from patency of the ductus arteriosus in which there is pulmonary hypertension In an infant with a large ductus the pulmonary hypertension is physiological Under such circumstances the heart is greatly enlarged, on auscultation a systolic and a mid diastolic murmur are audible, the electrocardiogram usually shows a balanced axis and evidence of "combined" hypertrophy Both cardiac catheterization and aortography will show that the predominant shunt is from left to right

In contrast to this, when the pulmonary hypertension is primary and the ductus arteriosus remains patent, the heart is small and the electrocardiogram shows evidence of right ventricular hypertrophy Although the aortogram may reveal a ductus, no blood will be seen to enter the lungs

Occasionally in older children and in young adults, it may be difficult to determine whether the ductus arteriosus or the pulmonary hypertension is primary In such instances the size of the heart, the volume of the shunt, the electrocardiographic findings, and above all the degree of pulmonary hypertension in relation to the systemic pressure must be carefully evaluated Evidence of right ventricular hypertrophy usually means long standing pulmonary hypertension If the difference between the systemic and pulmonary pressures is such that there is a continuous murmur, it is usually safe to close the ductus arteriosus regardless of the degree of pulmonary hypertension If, however, the pressure in the pulmonary artery so nearly approaches the systemic pressure that there is no murmur, the operative risk for closure of the ductus arteriosus in patients over six years of age is extremely high, nearly 25 per cent If operation is attempted, the pressure in the pulmonary artery and in the aorta should be taken at operation both before and after clamps are placed on the ductus arteriosus If, upon the occlusion of the ductus, the pulmonary pressure falls and the systemic

ple ligation of the ductus arteriosus will sufficiently decrease the load on the heart so that closure of the ventricular septal defect may not be necessary or even advisable

The problem presented by *patency of the ductus arteriosus combined with pulmonary hypertension* is discussed above and also in detail in Chapter XVIII, Section B The occurrence of *persistent patency of the ductus arteriosus with coarctation of the aorta* is also well known (see Chapter XXVII)

It is a curious fact that a patient with the left coronary artery arising anomalously from the pulmonary artery, fibro-elastosis, Fiedler's myocarditis, or glycogen-storage disease seldom has persistent patency of the ductus arteriosus Perhaps it is only the comparative rarity of these conditions Perhaps it is that, with the exception of the anomalous coronary artery, these conditions may be metabolic or infectious in etiology and not related to the genes which affect the development of the heart. Hence with a basically normal heart, the ductus undergoes normal obliteration

COMPLICATIONS

Subacute bacterial endocarditis is by far the most common complication As in all malformations, bacteria tend to lodge where the abnormal currents of blood impinge against the vessel wall Hence, in this malformation, the bacteria lodge on the pulmonary artery opposite the point of entrance of the ductus Occasionally the fungating mass develops at the arterial end of the ductus or in rare instances may extend through its entire length Inasmuch as the bacteria usually are caught at the pulmonary end, the first emboli are thrown off into the lesser circulation infarcts in the lungs and pneumonia are among the early manifestations, whereas petechiae and splenic infarcts appear late and a positive blood culture may be difficult to obtain

It is important to remember that the presence of the fungating mass may alter the physical findings When such a mass is located in the pulmonary artery, there may be obstruction to the pulmonary blood flow, thereby causing a functional pulmonary stenosis

Penicillin has proved so effective in the treatment of subacute bacterial endocarditis that medical treatment should always be instituted before an operation is considered If there is a prompt response to treatment, operation may be performed after four weeks of treatment prior to the final two weeks of therapy

It is also important to remember that Touroff and Vesell[12] showed that in a patient with subacute bacterial endocarditis simple ligation of the ductus ar

sus, the murmur is, however, more superficial and is maximal at a lower level, that is, in the third and fourth left interspaces close to the sternum The murmur is of maximal intensity slightly below the anatomical position of the ductus instead of above it The systolic murmur is often not as intense as that heard in a patent ductus arteriosus and the diastolic murmur is more intense

Other rare conditions which cause a continuous murmur must be considered if the continuous murmur is maximal over the lower portion of the precordium These conditions are discussed in the differential diagnosis of rupture of an aneurysm of the sinus of Valsalva (see Chapter xxv)

Conditions which cause aortic insufficiency or pulmonary insufficiency in infancy may be confused with patency of the ductus arteriosus These rare malformations are ordinarily of such a nature as to produce a systolic as well as a diastolic murmur, consequently the murmur may be mistaken for the continuous murmur of a patent ductus arteriosus Therefore in early infancy an aortogram may be indicated to differentiate these conditions

COMMONLY ASSOCIATED MALFORMATIONS

Inasmuch as the ductus arteriosus is a normal fetal pathway, persistent patency of the ductus arteriosus occurs with virtually every malformation As previously mentioned, in the presence of severe pulmonary stenosis or pulmonary atresia combined with a right to-left shunt at the ventricular or even at the auricular level, the patency of the ductus arteriosus increases the circulation to the lungs and is of benefit to the patient In such malformations, the ductus arteriosus should never be closed unless it is certain that the other abnormality can be corrected at the same time

The situation is quite different when the shunt is from left to right Under such circumstances the patency of the ductus arteriosus further increases the load placed on the heart Therefore closure of the ductus arteriosus is indicated, it is usually advisable to do so prior to the correction of the other malformation, because simple closure of the ductus arteriosus may so greatly relieve the load on the heart that further operation is not necessary

The occurrence of *a ventricular septal defect and persistent patency of the ductus arteriosus* is an excellent example of such a combination of anomalies If the patency of the ductus arteriosus is indicated by the presence of a continuous murmur, this in itself is positive proof that the pulmonary pressure is low Inasmuch as the combined load of the septal defect and the ductus arteriosus has not seriously elevated the pulmonary pressure, there is good reason to hope that sim

infection and must receive appropriate therapy Even after the successful closure of the ductus arteriosus it is probably a wise precaution for all patients to receive prophylactic therapy prior to dental extraction

Limitation of exercise is seldom necessary Most children limit their own activity If dyspnea is present, early operation is indicated If the operation is to be postponed to a more convenient season, the child should be permitted to live as normal a life as possible during the intervening period

Surgical treatment is highly satisfactory Either division of the ductus, as advocated by Gross,[0] or suture ligation, as recommended by Blalock,[1] gives excellent results

Suture ligation has the advantage that the operation is simple and short Division of the ductus is preferable if the ductus is large or too short to permit multiple sutures The risk of operation is remarkably low, approximately 2 per cent Indeed, the risk of surgery is as low as the risk of the subsequent danger of subacute bacterial endocarditis or cardiac failure The advantages of a normal heart are many It prevents the person from being heart conscious It should enable a person to get life insurance and increase the ease with which he can get employment It removes one possible hazard during pregnancy

INDICATIONS FOR OPERATION

Inasmuch as the risk of operation is slight and the advantages are great, operation is indicated for any child with the classic form of this malformation

If the diagnosis is made in an infant with a heart of normal size, it is usually wise to postpone operation at least until the patient is two years old, as the ductus arteriosus may still close of its own accord If, however, the heart is greatly enlarged and the infant suffers from respiratory distress or repeated respiratory infections, early operation is indicated

When the condition is discovered in infancy or in early childhood and the patient

desire [illegible]

tainly there is no necessity to perform the operation the instant the diagnosis is made The ideal age for operation is between three and eight years Operations can however be performed safely on infants and young adults

If the patient is over thirty years of age and has a heart of normal size and function and is leading a normal life, the only argument in favor of operation is the prevention of subacute bacterial endocarditis Since this complication is now curable closure of the ductus arteriosus for such a person is highly elective

teriosus sterilizes the blood stream provided the vegetations are limited to the ductus and do not involve the valves Therefore, if the infection does not respond promptly to medical treatment, early operation is indicated Delay only increases the danger of emboli, especially pulmonary infarction, and renders operation more difficult

Aneurysmal dilatation of the ductus arteriosus is a rare complication The first such case was reported by Hebb[13] in 1893, the next by Altschule[14] in 1937 In 1940 Graham[15] reported two additional cases An aneurysmal dilatation of the ductus arteriosus occurs only when the obliteration of the ductus is complete at the pulmonary end but incomplete at the aortic end Under such circumstances there is no longer an arteriovenous connection The signs are not those of patency of the ductus arteriosus but those of an aneurysm

Aneurysms of the pulmonary artery, both proximal and distal to the entrance of the ductus arteriosus, have been reported[16, 17] Dilatation of the pulmonary artery proximal to the ductus is one of the characteristic x ray findings in a patent ductus arteriosus If there is a congenital weakness of the wall of the pulmonary artery, aneurysmal dilatation may develop The formation of an aneurysm in the pulmonary artery is the exception, not the rule The rarity of these aneurysms indicates that they occur only when the pulmonary wall is weakened by disease or by some congenital abnormality

TREATMENT

Surgical closure of the ductus arteriosus was first reported by Gross and Hubbard[18] in 1939 That same year Bullock et al[19] reported eleven patients in whom the ductus arteriosus had been successfully closed These operations showed that in patients with persistent patency of the ductus arteriosus it was possible to restore the heart and the circulation to normal The operation is now performed with an extraordinarily low mortality rate, therefore operation is the treatment of choice

Medical treatment is indicated prior to surgery only if the patient is in cardiac failure or has subacute bacterial endocarditis

Prophylactic penicillin or chemotherapy is essential prior to dental extraction or tonsillectomy in a patient with patency of the ductus arteriosus Among the patients reported at the Second World Congress, over a period of fifteen years no instance of subacute bacterial endocarditis had been known to occur after successful closure of the ductus arteriosus The ductus arteriosus has been known to recanalize Under such circumstances the patient is again susceptible to bacterial

cardiogram showed inversion of the T waves in all three of the standard leads *Usually*, however, the electrocardiogram has returned to normal within three months

In patients with marked cardiac enlargement dilatation of the heart promptly disappears The disappearance of hypertrophy takes longer *Nevertheless*, once the load on the heart has been decreased, the heart will remain stationary in size, the chest will grow, and eventually the child will outgrow the cardiac enlargement During this period there is usually no necessity for limitation of activity

Postoperative complications are rare There may be mild hypertension immediately after operation this is, however, usually transitory and need cause no concern

Aphonia, due to injury to the recurrent laryngeal nerve, is also a rare complication Generally within a month the patient's voice returns

Recanalization of the ductus is probably the most serious complication Fortunately this is rare Curiously enough, in Crafoord's - experience, recanalization has occurred even after the division of the ductus, nevertheless, when the ductus recanalizes it opens into the pulmonary artery

Recanalization, if it occurs, usually happens within two weeks to two months after operation After this interval, complications from operation are rare

Once the patient has recovered from operation, the circulation is normal The individual may be permitted normal activity Men are accepted into the Armed Forces one year after successful operation

PROGNOSIS

The prognosis for a patient with a patent ductus arteriosus is good Many patients can lead long and active lives without operation Nevertheless, since the risk of operation is slight and it restores the heart and circulation to normal, operation is usually indicated After such a procedure the prognosis is excellent

SUMMARY

Persistent patency of the ductus arteriosus may be caused by German measles during the first trimester of pregnancy

The ductus arteriosus is a normal fetal pathway which usually closes shortly after birth Closure may, however, occasionally be delayed for several months Therefore in early infancy if the ductus arteriosus is normal in size and position, its patency should not be considered a malformation

Failure of the ductus arteriosus to undergo normal obliteration constitutes an

If surgery is recommended every time patency of the ductus arteriosus is suspected, some errors in diagnosis will inevitably be found at operation An aortic septal defect and a hemi truncus arteriosus are readily confused with patency of the ductus arteriosus Under such circumstances the chest should be closed and *nothing more attempted* without further study and consideration

The major contraindication for operation is the existence of another malformation of such a nature that the ductus is acting in a compensatory manner For example, with pulmonary stenosis or atresia, patency of the ductus arteriosus is of great benefit to the patient and therefore the ductus arteriosus should not be ligated For this reason, whenever any additional malformation is suspected, it is important to make certain that the oxygen saturation of the arterial blood is within normal limits and that the shunt is overwhelmingly from left to right Cyanosis and clubbing are usually indicative of a venous arterial shunt In the presence of such findings, unless it can be proved that the patient has a huge ductus and that the pulmonary blood flow is adequate without the shunting of blood from the aorta to the pulmonary artery, the ductus arteriosus should not be ligated Although mild pulmonary hypertension is an indication for operation, severe pulmonary hypertension is usually a contraindication, especially if the heart is small and the electrocardiogram shows evidence of marked right ventricular hypertrophy (see page 512 and Chapter xviii)

The benefits from operation are real and may be dramatic The continuous murmur disappears and the pulse pressure returns to normal The heart action becomes quiet Convalescence is remarkably rapid The patient is generally ready *for discharge from the hospital a week after operation* Exercise should, however, be *restricted for the ensuing month* Six weeks after operation it is safe for the patient to resume normal activity

If, however, the condition has been complicated by subacute bacterial endocarditis, the patient should have the benefit of a longer period of bed rest Even though the blood stream is instantly sterilized, it is *inconceivable that the entire* fungating mass will immediately disappear The patient should, therefore, have the benefit of additional rest until there is reasonable probability that the focus of infection has been eliminated Under such circumstances convalescence usually requires from two to three months

A functional murmur is usually audible over the base of the heart for many months after surgical closure of the ductus The vascular markings gradually diminish and over a period of months the electrocardiogram returns to normal In one patient, six months after the closure of a large patent ductus, the electro-

The standard leads of the electrocardiogram show a balanced axis and the unipolar precordial leads usually show evidence of combined hypertrophy

Cardiac catheterization shows an increased oxygen content in the blood in the pulmonary artery It may or may not be possible to pass the catheter through the ductus into the descending aorta

Aortography is probably the most useful test to determine patency of the ductus arteriosus in infants

Before the development of a continuous murmur the condition is to be suspected in an infant with a whipping pulse and a large heart and both a systolic and a diastolic murmur over the precordium, especially when these findings are combined with x ray evidence of increased vascularity of the lungs and electrocardiographic evidence of a balanced axis or of left axis deviation and combined hypertrophy

In children and young adults who show no cyanosis, the diagnosis is readily made by the finding of the characteristic continuous murmur over the pulmonary area

The condition requires differentiation from other malformations with increased pulmonary blood flow and from those with a continuous murmur In infancy the condition is most commonly confused with a large ventricular septal defect or an aortic septal defect In childhood it must be differentiated from a venous hum, an aortic septal defect, a truncus arteriosus with increased flow, or a hemi truncus arteriosus

Primary pulmonary hypertension in association with patency of the ductus arteriosus requires differentiation from patency of the ductus arteriosus with pulmonary hypertension secondary to it If there is a continuous murmur, closure of the ductus is usually well tolerated In older children and in young adults, in the absence of a continuous murmur, surgical closure of the ductus should be attempted only if the predominant shunt is from left to right and if at operation the pressure in the pulmonary artery drops after clamps are placed upon the ductus

In addition to the above conditions, persistent patency of the ductus arteriosus in adults must be differentiated from a large ventricular septal defect combined with aortic insufficiency, from rheumatic heart disease with aortic insufficiency and also from a rupture of an aneurysm of the sinus of Valsalva into the right auricle or the right ventricle and other rare conditions in which there is a continuous murmur

abnormality Persistent patency of the ductus arteriosus causes oxygenated blood to be shunted from the aorta into the pulmonary artery This increased volume of blood is returned to the left auricle and the left ventricle, hence these chambers are enlarged Because of the increased pulmonary blood flow, the work of the right ventricle is also increased, the right ventricle, too, is enlarged

The malformation is approximately twice as common in females as in males

The symptoms vary with the size of the ductus

In infancy and early childhood, if the ductus is small, even though the ductus arteriosus is patent there may be no clinical manifestations If, however, the ductus arteriosus is large, it may cause great difficulty

Failure to gain and stunting of growth may occur in infancy

Cyanosis seldom occurs, as the direction of the shunt is from left to right

The pulse pressure is usually wide

A continuous murmur over the pulmonary area is the most characteristic of all findings This murmur, however, is not present at birth and is seldom heard until the child is over two years of age Prior to the development of the continuous murmur, diagnosis is difficult but accurate diagnosis is only necessary in the presence of great cardiac enlargement

After the development of a continuous murmur, the malformation is one of the simplest to diagnose clinically The continuous murmur, known as a "machinery" murmur, is virtually diagnostic of the malformation It is best heard over the base of the heart and is maximal in the second left interspace beyond the border of cardiac dullness The murmur has a harsh, rasping quality with a systolic accentuation but is continuous throughout diastole The systolic element of the murmur is transmitted to the vessels of the neck and is usually audible posteriorly, high in the interscapular region The murmur is louder in the recumbent than in the erect position and increases in intensity with exercise Usually it is of sufficient intensity to be accompanied by a palpable thrill

The heart may or may not be enlarged The size of the heart is directly related to the size of the ductus and the volume of the shunt The shunt is always from left to right There is no cyanosis and no clubbing When the shunt is large, the pulse is of the Corrigan type, there are peripheral signs of aortic insufficiency

X ray and fluoroscopy show varying degrees of cardiac enlargement There is usually, but not always, prominence of the pulmonary conus The hilar markings are increased but the hilar pulsations are relatively slight

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Surgical closure of the ductus arteriosus restores the heart and circulation to normal In the presence of cardiac failure or subacute bacterial endocarditis, prior to surgery the patient should receive appropriate medical therapy

Without operation the condition is usually compatible with an active life for a number of years After successful surgery the prognosis is excellent

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FIGURE XXI-1 Aortic septal defect *Infant*

This defect measured 1.2 x 1.3 cm and lay 0.5 cm above the aortic valve

COURSE OF THE CIRCULATION

During fetal life the course of the circulation is unaltered The defect merely acts as one more pathway by which a balance between the two circulations may be maintained

After birth with the expansion of the lungs and the consequent fall in pulmonary pressure, blood will flow from the aorta to the pulmonary artery There is a great increase in the pulmonary blood flow and, furthermore, blood from the aorta is directed to the lungs under systemic pressure Consequently the lungs open up more slowly than is normal and the pulmonary resistance remains relatively high The systemic and pulmonary pressures are so nearly equal that in the early months of life some blood may flow from the pulmonary artery into the aorta The predominant shunt is, however, from left to right and the shunt increases as the pulmonary vascular bed gradually expands The course of the circulation is similar to that in persistent patency of the ductus arteriosus

The blood from the right auricle flows into the right ventricle and thence is pumped out through the pulmonary artery to the lungs, where it is oxygenated The oxygenated blood is returned in the normal manner to the left auricle and

CHAPTER XXI

AORTIC SEPTAL DEFECT

IN 1830 Elliotson[1] reported a case of a "defect in the septum which is common to the aorta and the pulmonary artery' This is probably the first report of an aortic septal defect In 1868 Fraentzel[2] reported a case of an aortic septal defect and commented that such an anomaly was an additional cause of an early diastolic murmur This patient was a woman who died of cardiac failure at the age of twenty five Since then numerous cases have been reported The malformation is probably more common than has hitherto been suspected Its clinical manifestations are so closely similar to those of persistent patency of the ductus arteriosus that, until surgical closure of a patent ductus was perfected, aortic septal defects were by and large overlooked or misdiagnosed

NATURE OF THE MALFORMATION

An aortic septal defect, more correctly described as a congenital aorticopulmonary fistula," results from failure in the formation of the base of the spiral septum which normally divides the truncus arteriosus into the aorta and the pulmonary artery Consequently the defect occurs in that portion of the aortic wall which is common to the pulmonary artery The defect lies close to the base of the aorta and may extend to the aortic cusps Usually, however, the defect lies from 1 to 5 mm above the base of the aorta and occasionally as much as 1 cm above the aortic valve The size of the defect varies from 5 mm to more than 1 5 cm in greatest diameter Figure XXI–1 is a photograph of a relatively large aorticopulmonary fistula In rare instances the defect lies appreciably higher up on the arch of the aorta as in the case reported by Fletcher et al[3] Such a defect must be associated with a gross abnormality in the formation of the aortic septum, because normally the pulmonary artery and the aorta are entirely separated from one another before the beginning of the aortic arch It is important to remember that, although the wall between the aorta and the pulmonary artery is defective, both great vessels are normally formed and each arises in the normal manner from its respective ventricle The malformation concerns the septum above the heart, the heart itself is normally formed

The pulse is strong and may have a whipping quality The pulse pressure is wide

CARDIAC FINDINGS

The heart is usually enlarged The *rate* is rapid The *murmurs* are closely similar to those of persistent patency of the ductus arteriosus In early infancy it is common to find a *systolic murmur* and a *thrill* over the precordium and often a blurred mid-diastolic murmur within the apex In addition there may be a gallop rhythm Occasionally the murmur is of maximal intensity more nearly in the mid line and at a slightly lower level than is usual in patency of the ductus arteriosus Under such circumstances the murmurs closely resemble those of a ventricular septal defect In many instances there is nothing which differentiates the murmurs from those produced by persistent patency of the ductus arteriosus or from that of a ventricular septal defect

As in a patent ductus arteriosus, when the lungs become sufficiently expanded so that the pulmonary pressure is significantly lower than the systemic pressure, a continuous murmur develops Since the pulmonary pressure generally drops more slowly than is usual with patency of the ductus arteriosus, the continuous murmur develops at a later age Neill and Mounsey[5] have emphasized that in an aortic septal defect the quality of the murmur may change over a period of years The author has followed one patient who initially had a murmur which was typical of a patent ductus arteriosus At operation no ductus was found but a thrill over the base of the pulmonary artery made it seem probable that there was an aortic septal defect At that time closure of an aortic septal defect was not possible and since then the family has not desired an operation The child now has a roaring systolic and diastolic murmur over the base which does not sound in the least like that of patency of the ductus arteriosus Nevertheless, in infants and young children the two conditions are extremely difficult to differentiate Martin[6] has found that the murmur of an aortic septal defect is frequently better heard in the right interscapular region than in the left

X RAY AND FLUOROSCOPIC FINDINGS

The heart is enlarged There is dilatation and hypertrophy of both ventricles and of the left auricle The pulmonary conus is full and the vascularity in the lungs is increased There may be a hilar dance Figure XXI-2 shows the contour of the heart in a patient whose aorticopulmonary fistula was successfully closed by Scott and Sabiston

thence it flows to the left ventricle and is pumped out from the left ventricle into the aorta As the lungs expand, the pressure in the pulmonary artery falls and some blood flows from the aorta through the defect to the pulmonary artery and re circulates through the lungs and is again returned to the left auricle Most of the blood in the aorta is directed to the body and returned in the normal manner by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again (see Diagram XXI–1)

PHYSIOLOGY OF THE MALFORMATION

Although the course of the circulation is similar to that of patency of the ductus arteriosus, the condition is more serious because the opening between the aorta and the pulmonary artery lies in the ascending part of the aorta and not in its descending part Reynolds and Light[4] have shown that, although the pressure is the same in the ascending and the descending aorta, in the ascending aorta there is both forward propulsion and lateral expansion, whereas in the descending aorta the expansile force has been lost and the pressure is almost entirely that of forward propulsion Consequently, in an aortic septal defect, blood is directed to the lungs under higher pressure than it is when the ductus arteriosus remains patent In the latter condition the increase in pulmonary pressure is primarily due to the volume of the pulmonary blood flow, whereas in an aortic septal defect the expansile force transmitted from the ascending aorta, as well as the volume of the pulmonary blood flow, elevates the pulmonary pressure For this reason, a large aortic septal defect is a more serious malformation than an equally large patent ductus arteriosus

CLINICAL FINDINGS

The clinical findings vary with the size of the defect, the age of the patient, and the relative pressure in the two circulations In many instances the findings closely simulate those of persistent patency of the ductus arteriosus

Failure to gain weight may be an outstanding complaint in early infancy

Dyspnea and *polypnea* are common Nevertheless, the infant does not suffer from attacks of paroxysmal dyspnea

Respiratory infections are of frequent occurrence

Cyanosis is usually absent, as the predominant shunt is from left to right Occasionally, when the septal defect is large and the pressure in the pulmonary artery is approximately the same as that in the aorta, sufficient venous blood may be shunted into the aorta to produce slight cyanosis

Diagram XXI-1

Aortic septal defect

The essential feature of this malformation is a defect in the wall common to the aorta and the pulmonary artery just above the aortic valves

The blood from the right auricle passes into the right ventricle and is pumped out through the pulmonary artery in the normal manner to the lungs The oxygenated blood from the lungs is returned by the pulmonary veins to the left auricle Thence it flows to the left ventricle, from which it is pumped into the aorta to the systemic circulation Inasmuch as the pressure in the aorta is higher than that in the pulmonary artery, some blood flows through the aorticopulmonary fistula into the pulmonary artery The oxygenated blood so shunted flows through the pulmonary artery to the lungs, where it re-circulates and is again returned to the left auricle There the cycle starts again

Thus in this malformation a shunt is established through the lesser circulation and the work of the left side of the heart is increased Furthermore, the blood shunted through the aortic septal defect is propelled to the lungs under the usual systemic pressure plus the expansile force which is present in the ascending aorta This force raises the pulmonary pressure, which in turn increases the pressure against which the right ventricle must work Therefore both ventricles are slightly hypertrophied and the left auricle is enlarged

Clinical diagnosis The size of the heart is determined by the size of the defect, which in turn determines both the volume of the shunt and the pressure under which the blood is shunted into the pulmonary artery Inasmuch as the shunt is from left to right there is no cyanosis and no clubbing

The physical findings are closely similar to those of persistent patency of the ductus arteriosus with the exception that the continuous murmur may be better heard to the right of the sternum than is usual in a patent ductus arteriosus and also may be audible in the right interscapular region Furthermore, the quality of the murmur may change over a period of years The electrocardiogram usually shows a balanced axis and combined hypertrophy

DIAGRAM XXI–1

Head and upper extremities

R Lung

S.V.C.

P.A.

Aorta

Pul A

L Lung

PV

Pulmonary Artery

Pul V

L A

R.A

Aortic Septal Defect

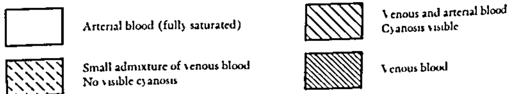
LV

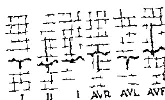
Descending Aorta

RV

I.V.C.

Trunk and lower extremities





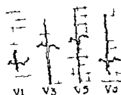


FIGURE XXI-3 Aortic septal defect (same patient as in Figure XXI-2) Infant

cannot be passed any further, it is more likely that it still lies in the pulmonary artery rather than in the aorta, as the blood ejected from the left ventricle tends to direct the catheter away from the defect in the aortic septum

Angiocardiography is of no aid in the diagnosis of this malformation

Aortography may be of great diagnostic aid The detection of dye in the lungs at the time of visualization of the aorta shows that there is a connection between the two circulations above the aortic valves The diagnosis can be made with assurance if the dye is seen to pass from the base of the aorta into the main pulmonary artery and both vessels are equally well opacified The diagnosis is further strengthened if at the same time the entire aortic arch and the descending aorta are so clearly delineated as to exclude the possibility of patency of the ductus arteriosus Thus an aortogram such as is shown in Figure XXI-4 may definitively establish the existence of an aortic septal defect but failure to obtain a characteristic picture does not exclude it

DIAGNOSIS

The diagnosis may be suspected when the findings are suggestive of a patent ductus arteriosus but the murmurs are maximal further to the right than is usual in that anomaly or when the intensity of the murmur or the degree of incapacity is greater than is to be expected from the size of the heart The observation that the quality of the murmur changes over a period of years aids in the diagnosis of an aortic septal defect Accurate diagnosis can seldom be made without recourse to special studies and even then the correct diagnosis is seldom established prior to operation If a patient has a continuous murmur over the pulmonary area and no ductus is found at operation, it is probable that he has an aortic septal defect



Figure xxi-2 Aortic septal defect (same patient as in Figures xxi-3 4) Infant

ELECTROCARDIOGRAPHIC FINDINGS

The standard leads usually show a balanced axis All the unipolar precordial leads show both a tall R and a deep S, indicative of hypertrophy of both ventricles Thus the electrocardiographic findings are closely similar to those associated with patency of the ductus arteriosus (see Figure xxi-3)

SPECIAL TESTS

The red blood cell count, the level of the available *hemoglobin,* and the *hematocrit reading* are within normal limits

Cardiac catheterization shows that the pulmonary artery arises from the right ventricle, that the main increase in the oxygen content of the blood occurs in the pulmonary artery, and, furthermore, that the pressure in the right ventricle and in the pulmonary artery approaches systemic pressure It is noteworthy that the pressure in the pulmonary artery tends to be higher in an aortic septal defect than in a ductus of the same size If the catheter passes through the aortic septal defect, the course of the catheter will follow that of the arch of the aorta and it will not swing abruptly into the descending aorta as it does when the ductus arteriosus is catheterized Only if the catheter is passed up into the neck vessels, as in the cases reported by Fletcher et al [3] and by D Heer and Van Nieuwenhuizen,[8] or down the descending aorta to a level below the diaphragm, is it positive proof that the aorta has been entered If the catheter loops anteriorly but

continuous murmur is, however, usually far more widely *transmitted* over the lungs (see Chapter XIV, Section B)

A truncus arteriosus with increased pulmonary blood flow may be confused, even at operation, with an aortic septal defect when the main pulmonary artery arises from an orifice at the base of the truncus The two conditions are quite different a true truncus is a single vessel which receives the blood from both ventricles and from which the pulmonary arteries arise, in contrast to this an aortic septal defect is a communication between two separate vessels (the aorta and the pulmonary artery) which arise from the left and right ventricles respectively The differentiation of these two conditions is extremely important, as closure of an aortic septal defect restores the heart and circulation to normal, whereas, if there is a truncus arteriosus, closure of the orifice of the pulmonary artery would completely cut off the circulation to the lungs and thereby be immediately fatal

A large ventricular septal defect with increased pulmonary blood flow and high pulmonary pressure is quite as readily confused with an aortic septal defect as with persistent patency of the ductus arteriosus Indeed, both of these malformations are differentiated from the ventricular septal defect by the same findings (see Chapter XX)

Rupture of an aneurysm from the sinus of Valsalva into the right ventricle and *other rare conditions which cause a continuous murmur* are differentiated from an aortic septal defect by the same methods as those used to differentiate them from patency of the ductus arteriosus (see Chapter XXV)

TREATMENT

Surgical closure of an aortic septal defect restores the heart and circulation to normal It is however, a more difficult operation than is the closure of a patent ductus arteriosus Indeed, in 1953 when Scott and Sabiston[7] reported the successful closure of an aorticopulmonary fistula in an infant of eighteen months, there had been only two other successful closures and one partial closure Since the advent of hypothermia and the use of a pump and oxygenator, the operation has become more readily and more safely accomplished Indeed, a number of successful closures[3, 9, 11] have been performed with these techniques

It is important to appreciate that the differential diagnosis between an aortic septal defect and a patent ductus arteriosus is extremely difficult and that errors in diagnosis are inevitable If such an error is made or, indeed, if at operation for patency of the ductus arteriosus no ductus is found, it is wise to close the

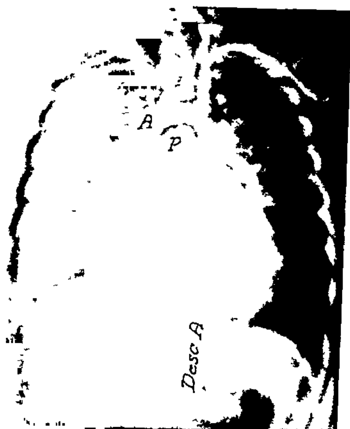


FIGURE XXI-4 Aortic septal defect (same patient as in Figure XXI-2) Infant

DIFFERENTIAL DIAGNOSIS

The condition is most commonly confused with patency of the ductus arteriosus and may require differentiation both from a truncus arteriosus with increased pulmonary blood flow and a hemi truncus arteriosus and also from a high ventricular septal defect with a large left to-right shunt, and occasionally from other rare conditions in which there is a continuous murmur

Patency of the ductus arteriosus is extremely difficult to differentiate from an aortic septal defect Occasionally the location of the murmur suggests the correct diagnosis Aortography may be of aid if the films are taken as the dye enters the lungs and if both the base of the aorta and the descending portion of the arch of the aorta are sharply delineated Cardiac catheterization is of value only if the catheter is passed through the defect and is seen as it courses through the arch of the aorta to the vessels of the neck or to the descending aorta

A hemi truncus arteriosus may be confused with an aortic septal defect The

considered The change in the quality of murmurs over a period of years is also suggestive of an aortic septal defect Even with special tests the correct diagnosis may not be made prior to surgery

The two outstanding conditions with which this malformation is confused are persistent patency of the ductus arteriosus and a truncus arteriosus in which the pulmonary artery arises from the base of the truncus

Surgical closure of an aortic septal defect is difficult but can be safely accomplished under direct vision with or without hypothermia

Prognosis varies with the size of the defect A small defect may be compatible with a long and active life A large defect may be corrected by surgery

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chest and reoperate at a later date unless the surgeon is prepared to switch to a more elaborate technique and the family have been advised of the seriousness of the operation Dissection around the base of the aorta will lead to the development of scar tissue and adhesions which will increase the difficulty of a future operation

PROGNOSIS

Although an aortic septal defect is a more serious abnormality than patency of the ductus arteriosus, a number of patients have lived to young adult life Successful closure of an aortic septal defect restores the heart and circulation to normal The operation, however, usually requires either hypothermia or a pump and oxygenator Nevertheless, if there is either severe pulmonary hypertension or considerable cardiac enlargement successful operation changes the prognosis from poor to excellent

SUMMARY

An aortic septal defect is a fistula between the aorta and the pulmonary artery just above the aortic valves The condition is frequently mistaken for patency of the ductus arteriosus

The course of the circulation is similar to that of a patent ductus arteriosus in that there is a left to-right shunt above the aortic valves The pressure in the pulmonary artery tends to be higher than that which occurs with a patent ductus arteriosus of similar size, because the blood is shunted into the pulmonary artery with the expansile force which exists in the ascending aorta

The clinical and the cardiac findings are closely similar to those of patency of the ductus arteriosus Occasionally the murmur is heard further to the right of the sternum than is usual in a patent ductus arteriosus, sometimes the murmur is relatively loud in the right interscapular region

Cardiac catheterization is of greater aid in the proof that the shunt is above the aortic valves than in the establishment of the diagnosis

Aortography may occasionally be of great value in that the dye appears simultaneously in the main pulmonary artery and at the base of the aorta and yet the arch of the aorta and the descending aorta are clearly delineated and no suggestion of a ductus is seen

The diagnosis may be extremely difficult If the findings are suggestive of a patent ductus arteriosus and if the murmur is maximal over the sternum or even to the right of the sternum, the possibility of an aortic septal defect should be

EMBRYOLOGY

Normally, at the time when the lung buds become prominent, there is but a single pulmonary vein which grows down to meet an outpouching in the posterior wall of the left auricle[1] This single pulmonary vein bifurcates as it passes to the lungs and the two main branches divide again and subsequently redivide as they extend into the lungs At first there is only a single pulmonary vein emptying into the left auricle As the left auricle expands, this vein becomes incorporated into it and subsequently the first and second branches of this vein become similarly incorporated, so that ultimately four pulmonary veins enter the left auricle

A total anomaly of the pulmonary venous return results if the primitive single pulmonary vein connects with the left common cardinal vein or with the left superior vena cava instead of with the outpouching from the left auricle Under such circumstances the left superior vena cava usually joins normally with the innominate vein and thus all the blood from the pulmonary veins drains into the right superior vena cava Occasionally the left common cardinal vein persists and extends downward as a persistent left superior vena cava which opens into the coronary sinus Under such circumstances the pulmonary venous return may open into this vein and thereby drain into the right auricle by way of the coronary sinus

Total or partial anomaly of the venous return may also occur if the auricular septum, as it forms, deviates to the left Under such circumstances it is obviously far easier for the right pulmonary veins to open into the right auricle and for the left pulmonary veins to enter the left auricle than it is for the reverse to happen For a detailed discussion of the embryology and pathology the reader is referred to the works of Neill,[1] McManus, Edwards,[3] and others

NATURE OF THE MALFORMATION

A single pulmonary vein may open into the superior vena cava or high up into the right auricle Such an anomaly causes little alteration in the stress and strain of the circulation The heart itself is normally formed, the ductus arteriosus undergoes normal obliteration, and the foramen ovale closes in the normal fashion

A partial anomaly of the venous return sufficient to cause symptoms means that two or more pulmonary veins open into the right auricle The commonest type of partial anomaly is that in which the pulmonary veins from the superior and middle lobes of the right lung join the right auricle close to the entrance of

CHAPTER XXII

ANOMALIES OF THE PULMONARY VENOUS RETURN

ANOMALIES of the pulmonary venous return are many and varied By far the most common and the most important of these anomalies are those in which some or all of the pulmonary veins enter either directly into the wall of the right auricle or into the superior vena cava These anomalies are presented in Section A Section B is concerned with the anomaly of the pulmonary venous return in which all the pulmonary veins unite to form a single vessel which pierces the diaphragm and enters the hepatic vein Section C (The Scimitar Syndrome) concerns the anomalous drainage of some or all of the right pulmonary veins into the inferior vena cava combined with an anomalous development of the right lung and extreme dextroposition of the heart

A *Anomalies of the Pulmonary Venous Return to the Right Auricle*

The anomalous entrance of some or all of the pulmonary veins into the right auricle is a common abnormality These anomalies have been confused both clinically and at autopsy with auricular septal defects The drainage of the pulmonary veins into the right auricle causes an excessive amount of oxygenated blood to be pumped around and around the pulmonary circulation The extent of the alteration in the circulation and the clinical syndrome vary with the number of pulmonary veins which follow the anomalous course and also with the structure of the auricular septum

One, some, or all of the pulmonary veins may drain into the right auricle When all the pulmonary veins drain into the right auricle they may do so through the superior vena cava, through the coronary sinus, or directly into the right auricle, in rare instances they may pierce the diaphragm and open into the hepatic vein (see Section B) or into the inferior vena cava (see Section C) The auricular septum may be intact, there may be only patency of the foramen ovale or there may be a gross defect in the auricular septum The latter is common Furthermore, the defect in the auricular septum acts in a compensatory manner, as it aids in the direction of blood to the systemic circulation

A brief review of the embryology of the pulmonary veins will help to clarify the nature of the malformation

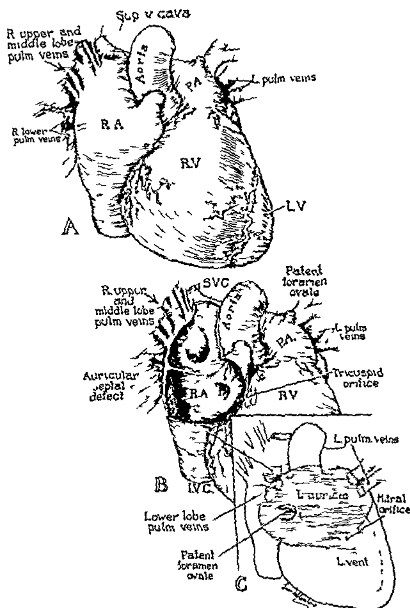


FIGURE XXII 2 Anomalous drainage of the right superior and middle pulmonary veins into the right auricle Child

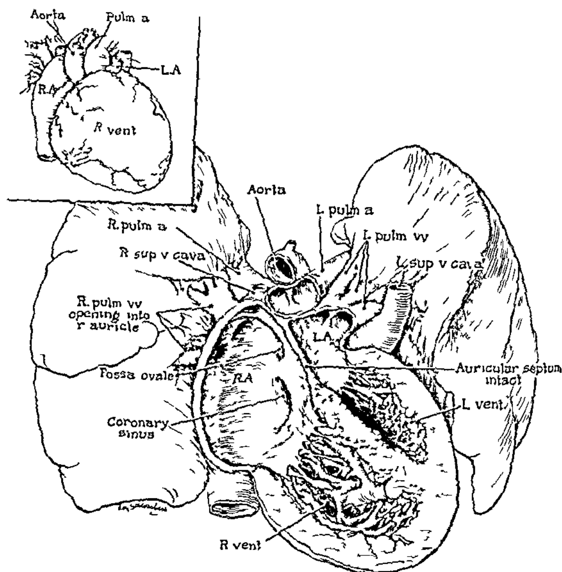


FIGURE XXII–1 Anomalous drainage of the right superior and middle pulmonary veins into the right auricle and an intact auricular septum (same patient as in Figure XXII–9) Child

the superior vena cava, as illustrated in Figure XXII–1 Sometimes these veins open into a quasi additional chamber in this region and adjacent to it there is a defect in the upper part of the auricular septum, as shown in Figure XXII–2 Occasionally this chamber is even more distinct and lies posterior to the right auricle and opens into the auricular wall close to the entrance of the superior vena cava Such an anomaly of the pulmonary venous return to the right auricle is not to be confused with a triauricular heart in which there is a partition between the upper and the lower portions of the left auricle (see Chapter XXX, *Section B*) The above mentioned anomaly is clearly a variant of the manner in which the

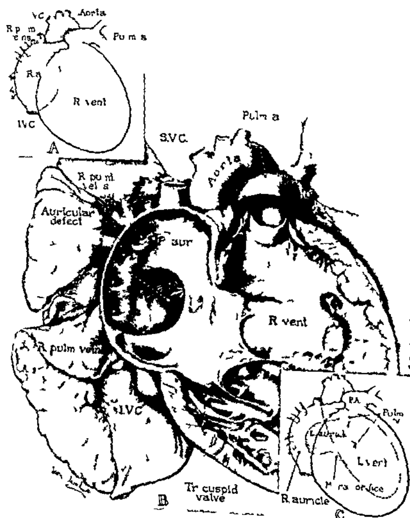


FIGURE XXII 3 Partial anomaly of the pulmonary venous return in which all the pulmonary veins from the right lung enter the right auricle (same patient as in Figure XXII 8) Adult

pulmonary veins enter the right auricle In other instances all pulmonary veins from the right lung may enter along the rim of the right auricle, as shown in Figure XXII–3 In all of the above instances the remainder of the pulmonary veins enter the left auricle in the normal fashion

A total anomaly of the venous return means that all the pulmonary veins drain into the right auricle The most common and the most distinctive of the anomalies of the pulmonary venous return occurs when all the pulmonary veins drain through a common channel into the left superior vena cava or the left common cardinal vein and thence through the innominate vein into the right superior vena cava,[4 7] as shown in Figures XXII–4 and 5 As previously mentioned, the common pulmonary vein may join with the persistent left superior vena cava to empty into the coronary sinus In rare instances the auricular septum may be shifted in such a manner that the pulmonary veins open directly into the right auricle

The most unusual of all the anomalies of the pulmonary venous return occurs when some or all of the pulmonary veins pierce the diaphragm and empty into the inferior vena cava either directly or through the hepatic vein The latter is an extremely serious malformation It produces a distinctive clinical picture and is therefore presented as a separate entity in Section B Section C gives the clinical syndrome of a partial drainage of the pulmonary veins into the inferior vena cava

When all the pulmonary veins empty into the right auricle, the left auricle has no connection with the lungs At autopsy it is possible to lift up the heart without disturbing the lungs This can readily be done when the pulmonary veins empty into the right superior vena cava When the pulmonary veins empty into the coronary sinus or into the posterior wall of the right auricle, it is possible to place one s hand between the left auricle and the lungs

In all three instances the structure of the heart is essentially the same The superior vena cava and the inferior vena cava open normally into the right auricle and in addition all the pulmonary veins empty their blood into this chamber The left auricle lacks its normal inflow tract It is a small chamber which can receive blood only from the right auricle Therefore, if the auricular septum is intact, the left auricle can receive blood only so long as the foramen ovale is physiologically patent

Since all the blood from both the systemic circulation and the pulmonary circulation enters the right auricle and the only blood that reaches the left auricle is the relatively small amount which passes through the foramen ovale, the right



FIGURE XXII–5 Anomalous drainage of all the pulmonary veins into a persistent left superior vena cava (same patient as in Figure XXII–4)

tive pressures in the two chambers tend to hold the foramen ovale open, and even though its patency is essential for life, the foramen ovale may tend to close in the normal manner This observation supports the finding reported by Barclay et al [8] that the closure of the foramen ovale depends more on the mechanical forces produced by the contraction of the auricles than upon the relative pressures in the two chambers

If there is no defect in the auricular septum, closure of the foramen ovale [illegible] der such circum

It is important to remember that in this anomaly the occurrence of a defect in the auricular septum is compensatory and may render the malformation compatible with life for many years Fortunately in a number of instances, when all the pulmonary veins drain into the superior vena cava, there is a gross defect in the auricular septum which enables blood to flow from the right auricle to the left auricle and thence to the left ventricle and the systemic circulation

When the pulmonary veins from the right lung drain into the right auricle

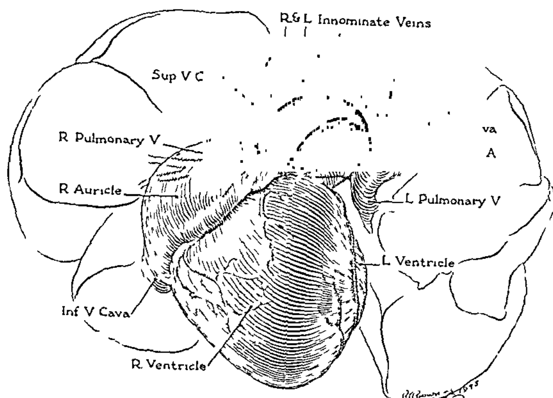


FIGURE XXII–4 Anomalous drainage of all the pulmonary veins into a persistent left superior vena cava and, by way of the left innominate vein, into the right superior vena cava and thence into the right auricle (same patient as in Figure XXII–5)

auricle and the right ventricle become greatly enlarged, the left auricle and the left ventricle remain small Underdevelopment of the left auricle and the left ventricle is inevitably far more common and more serious in a total anomaly of the venous return than it is in a partial anomaly, when some of the pulmonary veins return in the normal manner to the left auricle

In some instances the anomaly of the pulmonary venous return may be due to failure of the left side of the heart to develop Under such circumstances the left auricle and the left ventricle are abnormally small and the aorta is also hypoplastic

Total anomalies of the venous return are almost always associated with either a defect in the auricular septum or patency of the foramen ovale Indeed, if the auricular septum is intact, patency of the foramen ovale is essential for life When all or most of the pulmonary veins drain into the right auricle, the pressure in the right auricle is increased and that in the left auricle is abnormally low Consequently the relative pressures are such that they tend to hold the foramen ovale open Nevertheless, it is a striking fact that even though the rela

If, however, there is a total anomaly of the pulmonary venous return, the left auricle and the left ventricle may be diminutive This finding suggests that, although only a little blood normally circulates through the lungs and is returned to the left auricle, the volume of blood is sufficient to affect the development of the left auricle and the left ventricle When no blood is returned to the left side of the heart, during fetal life the major part of the systemic circulation is sustained by the blood pumped from the right auricle to the right ventricle and into the pulmonary artery and through the ductus arteriosus to the systemic circulation The left auricle and the left ventricle remain small On the other hand, it is possible that the failure of the left auricle to expand is the primary reason for the failure of the pulmonary vein to open into the left auricle Thus the underdevelopment of the left auricle and the left ventricle may cause this anomaly

After birth with the expansion of the lungs, the volume of blood which flows to the lungs is tremendously increased The course of the circulation varies, depending upon whether there is a partial or a total anomaly of the pulmonary venous return and whether or not there is a gross defect in the auricular septum

When but a single pulmonary vein opens into the superior vena cava although there is a slight increase in the volume of blood returned to the right [illegible] Under such circumstances, [illegible] the right auricle, where it a small volume of oxyg[illegible] mixes with the venous blood returned to the right auricle All the blood in the right auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs Most of the blood from the lungs is returned to the left auricle thence it flows to the left ventricle and is pumped out through the aorta to the body and returned in the normal manner by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again (see Diagram XXII-1)

When there is a partial anomaly of the pulmonary venous return some of the blood from the lungs is returned in the normal fashion to the left auricle and several pulmonary veins enter the right auricle In this anomaly there is usually a gross defect in the auricular septum Under such circumstances most of the blood from the right auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs, where it is oxygenated The blood which is returned in the normal fashion to the left auricle flows to the left ventricle thence it is pumped out through the aorta to the systemic circulation and is returned by the superior vena cava and the inferior vena cava to the right

and those from the left lung go to the left auricle, and there is also a gross defect in the auricular septum, the malformation is readily compatible with life for a number of years In contrast to this a partial anomaly of the pulmonary venous return combined with an intact auricular septum places such a severe strain on the right side of the heart that few patients survive beyond early childhood

COURSE OF THE CIRCULATION

During fetal life most of the blood which enters the left auricle normally comes from the right auricle and only a little blood is returned by the pulmonary veins to the left auricle Therefore, when there is a partial anomaly of the venous return, the course of the fetal circulation is not grossly altered The left auricle and the left ventricle receive virtually their normal quota of blood from the right auricle When the left side of the heart is normally formed, even a total anomaly of the venous return places no great strain on the fetal circulation At birth the heart is normal in size and shape (see Figure xxii–6)

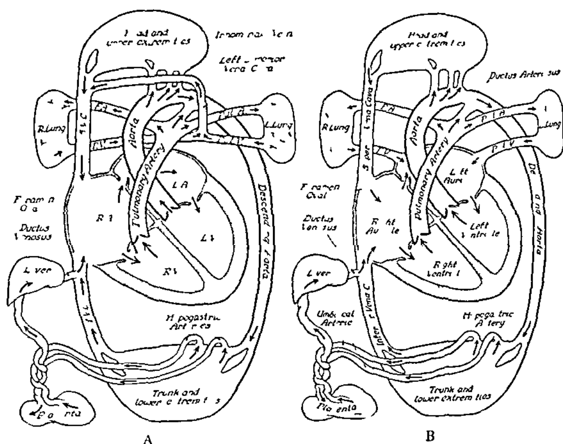


FIGURE XXII–6 Fetal circulation (A) Anomalous drainage of all the pulmonary veins into the right auricle by way of a persistent left superior vena cava and (B) normal heart

DIAGRAM XXII-1

Anomalous drainage of a single pulmonary vein into the right auricle

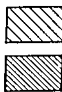
When a single pulmonary vein drains into the superior vena cava it causes little alteration in the circulation Although the oxygen content of the blood which enters the right auricle by the superior vena cava is a trifle higher than normal it becomes mixed with venous blood from the inferior vena cava this mixed venous blood flows into the right ventricle and is pumped out through the pulmonary artery to the lungs, where it is oxygenated Although a small volume of the oxygenated blood is returned by the anomalous pulmonary veins to the superior vena cava, most of the blood from the lungs is returned in the normal fashion to the left auricle thence it flows to the left ventricle and is pumped out through the aorta to the body The blood from the body is returned in the normal manner by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again

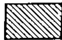
Clinical diagnosis is difficult Cardiac catheterization will show an increase in the oxygen content of the blood in the superior vena cava at the point of entrance of the pulmonary vein If the catheter is passed out into the lungs before it reaches the cardiac shadow it indicates that an anomalous pulmonary vein has been entered from the superior vena cava

Diagram xxii–1


Arterial blood (fully saturated)


Small admixture of venous blood
No visible cyanosis


Venous and arterial blood
Cyanosis visible


Venous blood

PHYSIOLOGY OF THE MALFORMATION

In this malformation the pulmonary blood flow is so tremendous that the pulmonary pressure is usually moderately elevated Although it usually does not reach the level of the systemic pressure, a systolic pressure of 60 to 80 mm of mercury in the pulmonary artery is frequently found Nevertheless, the tremendous pulmonary blood flow and the relatively small systemic blood flow cause a large volume of oxygenated blood to be mixed with a relatively small amount of venous blood, it follows that the oxygen saturation of the arterial blood is seldom reduced to the level which gives visible cyanosis Indeed, the oxygen saturation of the arterial blood is frequently between 97 and 99 per cent Moreover, the oxygen saturation of the blood directed to the lungs is equally high, hence the effective flow is low in spite of the enormous pulmonary blood flow

Occasionally, in early infancy, the pulmonary vascular bed opens up extremely slowly If this occurs, less blood is directed to the lungs and hence less oxygenated blood is returned to the right auricle Furthermore, the high pressure in the pulmonary artery raises the pressure in the right ventricle, which in turn raises the pressure in the right auricle, and consequently more venous blood is shunted through the foramen ovale to the systemic circulation Inasmuch as both factors increase the oxygen unsaturation of the arterial blood, the child may show cyanosis In addition, if there should be a collapse of the systemic circulation sufficient deoxygenation may occur in the peripheral tissues to produce visible cyanosis

The malformation places a tremendous strain on the right side of the heart The right auricle and the right ventricle are dilated and hypertrophied and the pulmonary blood flow is excessive, the left auricle and the left ventricle are spared and frequently are small and poorly developed chambers

CLINICAL FINDINGS

The appearance of the patient may be of diagnostic aid Children with this malformation may be unusually small for their age, Young adults may show the frail build and the poor physical development of the so-called *gracile habitus* The bones of such individuals are lighter than those of the average person and the skin has a semi translucent appearance and is of a fine, delicate texture The *onset of puberty* may be delayed These findings, which were first described as characteristic of an auricular septal defect,[9] are seen when the pulmonary blood flow is of such magnitude that the systemic circulation is starved For this reason they are frequently seen in patients with a *partial* anomaly of the venous return

auricle Since there is an anomaly in the pulmonary venous return, some blood from the lungs is *returned to the right auricle* Consequently the left auricle receives less than its normal quota of blood and the right auricle receives more than its normal quota Therefore the pressure in the right auricle is greater than that in the left If there is a defect in the auricular septum, some blood is shunted from the right auricle to the left auricle and the remainder flows into the right ventricle So the cycle continues (see Diagram XXII-2) The defect in the auricular septum increases the volume of blood directed to the systemic circulation and prevents the direction of an ever increasing volume of blood to the lungs The left ventricle, however, receives less than its normal quota of blood and the pulmonary circulation is excessive

When the auricular septum is intact, no blood can be shunted from the right auricle to the left auricle Therefore all the blood from the right auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs, where it is oxygenated Part of the blood from the lungs is returned to the left auricle, thence it flows to the left ventricle and is pumped out through the aorta to the systemic circulation and is returned in the normal manner by the superior vena cava and the inferior vena cava to the right auricle The right auricle also receives the oxygenated blood returned by the anomalous pulmonary veins, this mixture of venous and arterial blood flows into the right ventricle There the cycle starts again Thus, with each cardiac cycle, all the blood which is pumped out from the left ventricle and some of the blood which is pumped out from the right ventricle is returned to the right side of the heart Consequently the right auricle and the right ventricle are greatly enlarged The course of the circulation is shown in Diagram XXII-3

When there is a total anomaly of the venous return, all the pulmonary veins drain into the right auricle In order for the malformation to be compatible with life there must be a gross defect in the auricular septum or the foramen ovale must be held open by the high pressure in the right auricle Under such circumstances, although a large volume of blood from the right auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs, some blood flows through the auricular defect to the left auricle and thence to the left ventricle and is pumped out through the aorta to the systemic circulation The blood from the body is returned in the normal manner by the superior vena cava and the inferior vena cava to the right auricle In addition, all the oxygenated blood in the lesser circulation is returned by the pulmonary veins to the right auricle There the cycle starts again, as shown in Diagram XXII-4

DIAGRAM XXII–2

Partial anomaly of the pulmonary venous return combined with a gross auricular septal defect

The essential feature of this anomaly is that some of the pulmonary veins drain into the right auricle and there is also a gross defect in the auricular septum

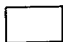
Inasmuch as some of the pulmonary veins drain into the right auricle, the pressure in the right auricle is increased and that in the left auricle is decreased and some blood from the right auricle flows into the left auricle Most of the blood in the right auricle flov [illegible] and is pumped out through the pulmonary artery to the lun [illegible]
au [illegible]
to [illegible]
right auricle to the left auricle This blood flows into the left ventricle and is pumped out through the aorta to the systemic circulation and returned by the superior and inferior venae cavae to the right auricle There it meets the blood returned by the pulmonary veins to the right auricle Thus the right auricle receives a mixture of oxygenated and venous blood Some of this blood flows to the left auricle through the tricuspid valve to the right ventricle So the cycle continues

Inasmuch as a large volume of blood is returned by the pulmonary veins to the right auricle the oxygen saturation of the blood in the right auricle is usually above 90 per cent Although a small amount of the mixed venous and arterial blood flows to the left auricle when it is combined with the blood returned in the normal manner by the pulmonary veins to the left auricle the blood in the left auricle and the left ventricle is even better saturated than that in the right auricle Indeed, the oxygen saturation of the blood in the femoral artery is usually from 97 to 99 per cent

Clinical diagnosis The patient has a frail build but shows no cyanosis The heart is enlarged and there is left sided chest deformity Murmurs are variable but usually a precordial systolic murmur and a reduplicated second heart sound are audible over the pulmonary area The x ray shows cardiac enlargement a full pulmonary conus, and increased vascularity The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy

The diagnosis should always be entertained if there is evidence of a gross defect in the auricular septum combined with increasing cardiac enlargement or if the oxygen saturation of the blood in the right auricle reaches 90 per cent or above

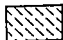
DIAGRAM XXII-2



Arterial blood (fully saturated)



Venous and arterial blood
Cyanosis visible



Small admixture of venous blood
No visible cyanosis



Venous blood

Diagram XXII–3

Partial anomaly of the pulmonary venous return combined with an intact auricular septum

The essential feature of this anomaly is that some of the pulmonary veins open into the right auricle and the remainder of the pulmonary veins enter the left auricle in the normal fashion and the auricular septum is intact

The blood from the right auricle flows into the right ventricle and is pumped out by way of the pulmonary artery to the lungs Part of the blood from the lungs is returned by the anomalous pulmonary veins to the right auricle and the remainder is returned in the normal fashion to the left auricle The blood from the left auricle flows into the left ventricle and is pumped out by way of the aorta to the systemic circulation The bloo [illegible] circulation is returned by the superior and inferior venae cavae to [illegible]
systemic circu [illegible]
left auricle receives only part of the blood from the pulmonary circulation The consequence is that the right auricle the right ventricle and the pulmonary circulation receive more than their normal quota of blood whereas the left auricle the left ventricle and the systemic circulation receive less than their normal quota of blood When all the pulmonary veins from the right lung drain into the right auricle and the auricular septum is intact the increased volume of blood returned to the right auricle places a great strain on the right side of the heart Inasmuch as part of the left ventricular output is returned to the right side of the heart the malformation places an ever increasing load on the right auricle and the right ventricle which ultimately leads to right sided cardiac failure Although the volume of blood which reaches the systemic circulation is reduced the blood is fully oxygenated

Clinical diagnosis The patient is dyspneic and has left sided chest deformity but he shows no cyanosis The heart is enlarged to the right and to the left There is fullness of the pulmonary conus and there are increased hilar markings Usually there is a systolic murmur and a low pitched mid diastolic murmur combined with accentuation of the second sound over the pulmonary area The electrocardiogram shows evidence of severe right sided cardiac strain Cardiac catheterization shows a marked increase in the oxygen content of the blood in the right auricle but it is impossible to pass the catheter into the left auricle

Diagram XXII-3

Head and upper extremities
R Lung
L Lung
P.A.
Pul A
Pul V
S.V.C.
Aorta
Pulmonary Artery
L A
R A
L V
R V
Descending Aorta
I.V.C.
Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible

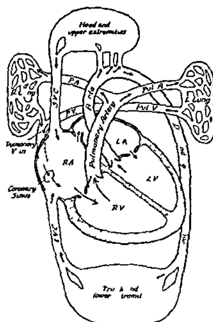

Small admixture of venous blood
No visible cyanosis


Venous blood

DIAGRAM XXII-4

Anomalous drainage of all the pulmonary veins into the right auricle by way of the superior vena cava

In this malformation all the pulmonary veins drain into a persistent left superior vena cava and thence into the innominate vein and via the right superior vena cava, into the right auricle. In addition, the foramen ovale is patent



Pulmonary veins enter the right auricle through the coronary sinus

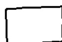
The greater part of the blood in the right auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs All the oxygenated blood from the lungs is returned by way of the left superior vena cava and the innominate vein to the right superior vena cava and thence to the right auricle Thus the right auricle receives both fully oxygenated and venous blood No blood is returned directly to the left auricle The pressure in the left auricle is low and some blood flows from the right auricle to the left auricle Thence the blood flows to the left ventricle and is pumped out through the aorta to the systemic circulation The blood from the body is returned by the superior and inferior venae cavae to the right auricle There the cycle starts again

As the foramen ovale closes more and more blood is pumped around the pulmonary circulation and the blood supply to the systemic circulation becomes progressively more inadequate Nevertheless since a large volume of oxygenated blood is mixed with a small volume of venous blood there is no visible cyanosis

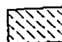
Clinical diagnosis There is progressive right sided cardiac enlargement but no cyanosis until the terminal collapse of the circulation The left superior vena cava and the large innominate vein cause the shadow at the base of the heart to be abnormally wide In children and young adults the wide mediastinal shadow gives the appearance of a snowman with his head off center A faint continuous murmur is sometimes audible high up to the right of the sternum The electrocardiogram shows a pronounced right axis deviation and evidence of right ventricular hypertrophy

When the pulmonary veins enter the right auricle through the coronary sinus (as shown in the insert) or enter directly into the posterior wall of the right auricle the course of the circulation within the heart is essentially the same as that given above. The shadow at the base of the heart is however not abnormally wide

Diagram XXII-4


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

A partial anomaly of the pulmonary venous return combined with a defect in the auricular septum also causes enlargement of the right side of the heart but the enlargement is not as extreme as that which occurs when all the pulmonary [illegible] the right auricle The contour of the heart is closely similar to [illegible]

years the malformation [illegible]

A partial anomaly of the pulmonary venous return combined *with an intact* auricular septum causes rapid cardiac enlargement Such children usually develop intractable cardiac failure by three years of age

The greatest cardiac enlargement occurs when a total anomaly of the pulmonary venous return is combined with a well formed auricular septum in which there is only patency of the foramen ovale Under such circumstances the malformation places an ever increasing strain on the right auricle and the right ventricle Indeed enormous cardiac enlargement may occur *within the first six* months of life The cardiac dullness to the right of the sternum is increased and that on the left side may extend to the axilla Even if there is a gross defect in the auricular septum, a total anomaly of the pulmonary venous return almost always causes cardiac enlargement at an early age There are, however, a few exceptions

The pulmonic second sound is usually accentuated

Murmurs are extremely variable In early infancy there may be *no murmur* In young children a *precordial systolic murmur* similar to or even harsher than that heard in an auricular septal defect is of common occurrence This murmur is probably produced by the flow of blood from the right auricle through a relatively small opening in the auricular septum into the left auricle The maximum flow of blood occurs in early diastole, *when the pressure within the* auricles is low and the blood rushes from the superior vena cava into the right auricle and thence directly through the foramen ovale into the left auricle Auricular diastole occurs *in early ventricular systole* Therefore a murmur of this origin would be systolic in time Once the auricles have filled, the flow of blood from the auricles to the ventricles causes no *greater disturbance than it* does in the normal heart Regardless of the validity of this explanation, the murmur over the precordium is systolic in time

The location of the systolic murmur is variable It may be maximal over the base but usually it is maximal in the third or fourth left interspace, midway between the sternum *and the apex*

The quality of the systolic murmur is extremely variable Indeed, the occurrence of a moderately loud systolic murmur which is too harsh for a functional

Left-sided chest deformity is the rule because of the great right sided cardiac enlargement

Dyspnea and easy fatigue on exertion are the rule Although the arterial blood is nearly fully oxygenated, the systemic blood flow is meager Furthermore, since the blood from the right auricle flows more readily into the right ventricle than it does into the left auricle and the left ventricle, the systemic blood flow is not readily increased by exercise These children seldom can keep up with their companions at play

Cyanosis is usually absent Although there is a complete admixture of venous and arterial blood in the right auricle, an excessive amount of oxygenated blood returned from the lungs is mixed with the relatively small amount of venous blood returned by the superior and inferior venae cavae, consequently the oxygen content of the blood in the right auricle is excessively high Hence the blood which reaches the systemic circulation is correspondingly well oxygenated Although the shunt is from right to left, cyanosis is usually absent until the terminal collapse of the circulation

Occasionally an infant or a child with this malformation may show persistent cyanosis which dates from birth This may occur if the pulmonary pressure is so high that the pulmonary vascular bed fails to expand in the normal manner Consequently less blood flows to the lungs and more blood flows to the body Under such circumstances the mixture of venous and arterial blood may be such as to produce visible cyanosis

Pulmonary congestion is the rule, it is due to the excessive pulmonary blood flow Furthermore, as the heart becomes greatly enlarged, it compresses the lower left lobe of the lung In infants the condition may be mistaken for pneumonia, as cyanosis is minimal, certainly not greater than is consistent with an extensive pneumonia, and the area of cardiac dullness may extend over the greater part of the left chest In addition, the enormous pulmonary blood flow renders these patients susceptible to pulmonary infections Slight respiratory infections frequently lead to bronchitis and bronchopneumonia

The liver is generally enlarged and extends several finger breadths below the costal margin Pulsations at the margin of the liver may or may not be demonstrable, depending on the strength of the cardiac contractions

Edema of the extremities is a late manifestation of cardiac failure

CARDIAC FINDINGS

The heart is enlarged The enlargement primarily involves the right auricle, the right ventricle, and the pulmonary artery and its branches

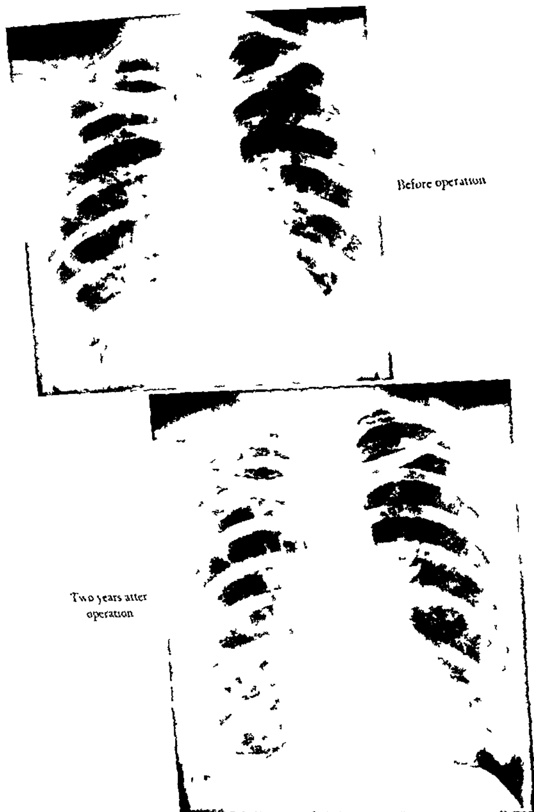


FIGURE XXII-7 Partial anomaly of the pulmonary venous return and an auricular septal defect Adult

murmur, but not as rasping as is usual in a ventricular septal defect, is always suggestive of an anomaly of the pulmonary venous return

A mid diastolic murmur or *a gallop rhythm* is frequently heard within the apex This is the murmur which is so commonly heard in the presence of great cardiac enlargement and cardiac failure

An apical systolic murmur which is transmitted toward the axilla is rarely heard in an anomaly of the pulmonary venous return, its occurrence is strongly suggestive that the auricular septum is intact

A soft continuous murmur is frequently heard high up to the right of the sternum when all the pulmonary veins drain into the superior vena cava This murmur is in all probability due to the currents of blood set up by the abnormally large volume of blood returned through the innominate vein to the superior vena cava

Cardiac arrhythmias may occur Broadly speaking, arrhythmias are likely to occur with enormous enlargement of one or both auricles Therefore it is not surprising that, in this malformation, paroxysmal tachycardia and excessive sinus tachycardia are relatively common Indeed, it may be the excessively rapid heart rate which brings the patient to the doctor

Cardiac failure although it does not invariably occur, is relatively common If there is evidence of progressive cardiac enlargement, sooner or later the condition leads to decompensation, with great engorgement of the liver Pulmonary congestion and rales in the lungs also occur because of the tremendous pulmonary blood flow

If cardiac failure has been caused by an excessive tachycardia, slowing of the heart rate by digitalis or other appropriate drugs will lead to temporary clinical improvement If, in spite of this, the heart continues to enlarge, the condition ultimately leads to intractable cardiac failure and death

X RAY AND FLUOROSCOPIC FINDINGS

The x ray and fluoroscopic findings vary with the extent of the anomaly of the pulmonary venous return

A partial anomaly of the pulmonary venous return associated with a gross defect in the auricular septum causes slight cardiac enlargement both to the right and to the left of the sternum, fullness of the pulmonary conus, and increased vascular markings which show a conspicuous hilar dance (see Figure XXII–7) Thus the x ray and fluoroscopic findings are closely similar to those produced by an auricular septal defect of the ostium secundum type (compare with Figure

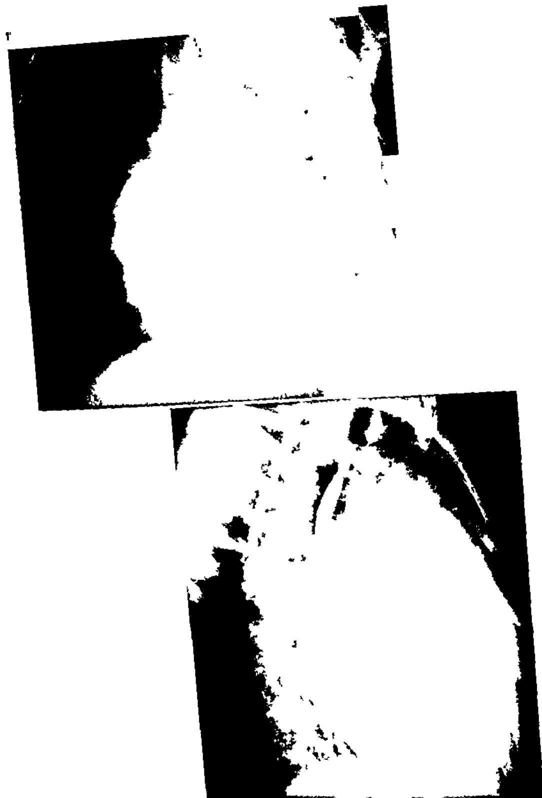


FIGURE XXII-8 Anomalous drainage of all the pulmonary veins from the right lung into the right auricle (same patient as in Figure XXII-3) Adult

xxiii–10) Furthermore, it is important to remember that a hilar dance is often more pronounced in this malformation than in an auricular septal defect

Examination in the left anterior oblique position confirms the fact that the great enlargement is due mainly to the increased size of the right auricle and the right ventricle In the right anterior oblique position there may occasionally be backward displacement of the esophagus This occurs when the greatly enlarged right auricle displaces the small left auricle further to the left than is usual and consequently the right auricle has pushed the esophagus backward (see Figure xxii–8) This finding is the exception, generally the esophagram is normal

A partial anomaly of the pulmonary venous return with an intact auricular septum leads to great enlargement of the right auricle and the right ventricle and excessive pulmonary congestion (see Figure xxii–9)

The drainage of all of the pulmonary veins into the right auricle by way of the superior vena cava causes the heart to assume a characteristic contour Under such circumstances not only is the right superior vena cava greatly dilated, but the shadow in the superior mediastinum to the left of the sternum is abnormally wide at the point where the pulmonary veins join the left superior vena cava

In young infants successive x ray films obtained over a period of time may show evidence of progressive cardiac enlargement Inasmuch as the strain on the heart becomes progressively greater as the foramen ovale tends to become sealed, the rate of cardiac enlargement becomes more rapid as the infant grows Between the ages of three and six months the heart enlarges very rapidly The rate of enlargement is so rapid that an increase in the size of the heart can usually be demonstrated within a period of weeks Terminally the cardiac shadow appears to fill the entire left chest Indeed, it may require careful examination to ascertain whether all of the shadow is cardiac in origin (see Figure xxii–10) In this instance, the infant was fluoroscoped but his condition was too critical to permit x rays

The sequence of events is quite different when there is a gross defect in the auricular septum, as the presence of the defect greatly relieves the strain on the right side of the heart and thus renders the condition compatible with life for a number of years

In older children and in young adults the contour of the heart becomes even more distinctive As the heart assumes a more nearly vertical position in the chest, the shadow of the superior mediastinum becomes narrower The heart and the superior mediastinum have the appearance of a snowman with the head slightly off center and to the right The shadow of the head is cast by the

anomalous pulmonary veins as they course from the junction of the left superior vena cava and the innominate vein to enter the right superior vena cava The body of the snowman is composed of the greatly dilated pulmonary artery, combined with enlargement of the right auricle and the right ventricle (see Figure XXII–11)

In addition, the lungs appear to be excessively vascular The vascular markings are widespread and extend far out into the lungs Usually there is a conspicuous hilar dance These shadows are even more diffuse than are those seen in patients with a gross defect in the auricular septum

In the left anterior-oblique position the heart frequently is seen to be flattened against the anterior chest wall and the enlargement of the right ventricle is so great that the left ventricle may be displaced backward Upon deep inspiration it may be possible to visualize the ventricular groove, thus confirming the fact that the enlargement is mainly right ventricular in origin

In the right anterior-oblique position the right ventricle appears to fill the entire chest Usually the esophagram is normal

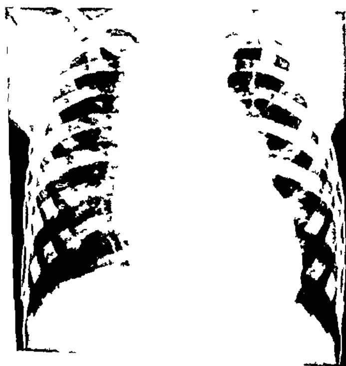


FIGURE XXII–11 Anomalous drainage of all the pulmonary veins into the right auricle by way of a persistent left superior vena cava Child



FIGURE XXII-9 Partial anomaly of the pulmonary venous return and an intact auricular septum (same patient as in Figure XXII-1) Child

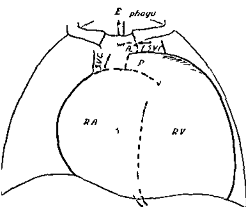


FIGURE XXII-10 Anomalous drainage of all the pulmonary veins into the superior vena cava Infant

anomalous pulmonary veins as they course from the junction of the left superior vena cava and the innominate vein to enter the right superior vena cava The body of the snowman is composed of the greatly dilated pulmonary artery, combined with enlargement of the right auricle and the right ventricle (see Figure XXII-11)

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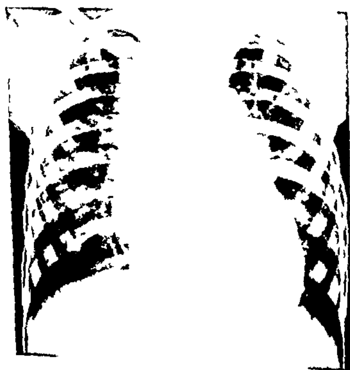


FIGURE XXII-11 Anomalous drainage of all the pulmonary veins into the right auricle by way of a persistent left superior vena cava Child

When the pulmonary veins drain into the coronary sinus or directly into the right auricle, the superior mediastinum remains clear There is, however, great enlargement of the right auricle Indeed, the right auricle extends nearly as far to the right as the right ventricle does to the left It is common to see a bulge in the upper part of the right auricle, as shown in Figure XXII–12

ELECTROCARDIOGRAPHIC FINDINGS

The standard leads show a right axis deviation and the unipolar precordial leads show evidence of right ventricular hypertrophy, which may be extreme The P waves may be abnormally high but are seldom notched (see Figure XXII–13)

When a partial anomaly of the pulmonary venous return is combined with an intact auricular septum, the electrocardiogram not only shows a right axis deviation and evidence of right ventricular hypertrophy in V_1 but also shows a tall R wave as well as a deep S wave in V_6

SPECIAL TESTS

The red blood cell count the amount of available hemoglobin, and the level of the hematocrit are generally within normal limits There may be a slight, but not severe, anemia

The oxygen saturation of the arterial blood in most instances is surprisingly high Although there is complete admixture of venous and arterial blood, the volume of the pulmonary blood flow is so great that the oxygen saturation of the blood in the right auricle is usually above 90 per cent, in many instances the mixed venous and arterial blood is from 96 to 98 per cent saturated The author has seen one patient whose blood appeared to be fully saturated in all four chambers of the heart

Cardiac catheterization is often of aid in the diagnosis of anomalies of the pulmonary venous return

If the catheter can be passed directly into a pulmonary vein and out to the lung before it reaches the cardiac shadow, it offers clear proof that one of the pulmonary veins opens into the superior vena cava

If a single pulmonary vein opens into the right auricle, the passage of the catheter into the pulmonary vein does not give conclusive evidence of the anomaly, as it is difficult to be certain whether the catheter entered the pulmonary vein directly from the right auricle or whether it slipped through the foramen

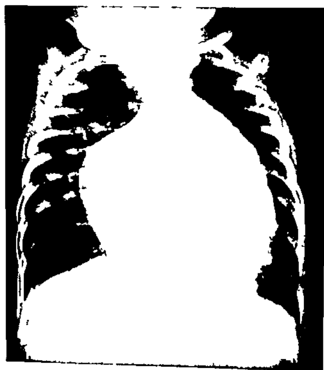


Figure xxii–12 Anomalous drainage of all the pulmonary veins into the right auricle by way of the coronary sinus Infant

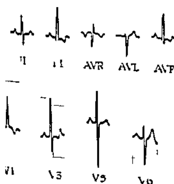


Figure xxii–13 Anomalous drainage of all the pulmonary veins into the superior vena cava

ovale or through an auricular defect and entered a normally placed pulmonary vein

Regardless of whether or not a pulmonary vein is *entered*, there is always an increase in the oxygen content of the blood in the right auricle as compared with that of a sample taken high in the *superior* vena cava The oxygen content of the blood in the right ventricle and in the pulmonary artery is also higher than that in either of the venae cavae but is not always as high as that in the *right* auricle

When one or more of the pulmonary veins enter the right auricle, the sample of blood taken close to the point of entrance of the vein will show a sharp increase in the oxygen *saturation* as compared with the other samples Frequently such a sample is found to be 97 or 98 per cent saturated Such an abrupt rise to nearly *full* saturation always suggests an anomaly of the pulmonary venous return

When the right superior and middle pulmonary veins enter the *right* auricle close to the entrance of the superior vena cava, the increase in the oxygen content of the blood is found high up in the right auricle If, however, all the samples taken from the right auricle are abnormally high, the catheterization findings may be similar to those produced by a gross defect in the auricular septum (see Chapter XXIII, Section B)

A total anomaly of the venous return in which all the pulmonary veins drain through the innominate vein into the right superior vena cava is readily confirmed by cardiac catheterization, provided the first sample of blood is taken high up in the right superior vena cava or in the brachial vein Under such circumstances a sample of blood taken from the area of the dilatation in the superior vena cava will show a marked increase in oxygen content as compared with a sample taken high up in the vein Indeed, the sample taken from the superior vena cava near the right auricle or taken from the innominate vein may be nearly fully saturated, that is, 97 or 98 per cent saturated The oxygen content of the blood in this sample will be the same as that taken from the right auricle, the right ventricle, and the main pulmonary artery Furthermore, the oxygen content of the blood in the femoral artery will be the same as that in the right auricle, as all the blood which reaches the left side of the heart comes from the right auricle

Indeed, the finding of the same oxygen content of the blood in all chambers of the heart which are entered and in the pulmonary artery and the aorta is always strongly suggestive of a total anomaly of the venous return, even if the blood is not nearly fully saturated

If, however, there is a marked increase in the oxygen saturation of the blood in the right auricle, right ventricle, and pulmonary artery to 93 per cent or higher, but the saturation on the right side is not as *high as that on the left*, it is strong presumptive evidence of a partial anomaly of the venous return The fact that one or more of the pulmonary veins open directly into the left auricle raises still further the oxygen content of the blood in the systemic circulation Nevertheless, it is important to bear in mind that when the oxygen content of the blood is extremely high there can be only slight variation in the several samples, these may be true differences but they are usually within the limits of experimental error

The pressure in the pulmonary artery in most anomalies of the pulmonary venous return is abnormally high The systolic pressure is frequently approximately 60 mm of mercury but it is usually not equal to the systemic pressure Occasionally the systolic pressure in the pulmonary artery may be as low as 40 mm of mercury

In brief, cardiac catheterization is usually of diagnostic aid, but only when the pulmonary veins drain into the superior vena cava does it offer positive proof of the anomaly

Angiocardiography is often of greater aid in the diagnosis of anomalies of the pulmonary venous return than is cardiac catheterization

When two or more of the pulmonary veins pour their blood high up into the right auricle it is usually possible to see a filling defect in the angiocardiogram where the blood from the pulmonary veins displaces the dye, as shown in the upper illustration of Figure XXII–14, or dye may be seen passing from the right auricle into one of the pulmonary veins, as shown in the lower illustration of Figure XXII–14

When the pulmonary veins drain into the right auricle at the junction with the superior vena cava there is frequently a defect in the auricular septum at this point Under such circumstances an angiocardiogram taken in the lateral position may show the dye passing into both auricles (see Figure XXII–15)

When all the pulmonary veins drain into the superior vena cava angiocardiography clearly shows the nature of the wide mediastinal shadow Occasionally, when these shadows are relatively inconspicuous in the x ray film, they may be clearly demonstrated by angiocardiography This is notably true in infants (compare the upper x ray of Figure XXII–16 with the lower angiogram of Figure XXII–17)

Early visualization of the aorta and the great vessels is far more common and

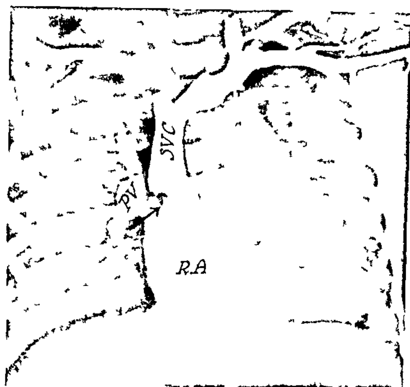




FIGURE XXII-14 Anomalous drainage of the upper pulmonary veins into the right auricle Child

Upper angiocardiogram shows the filling defect as blood enters the right auricle from the lung Lower angiocardiogram shows dye passing from the right auricle into the pulmonary vein

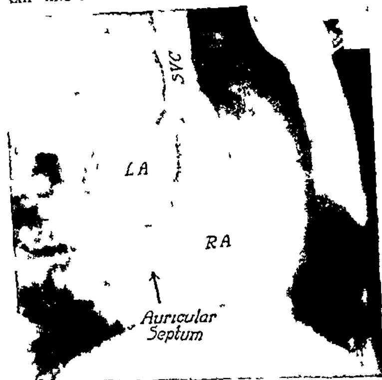


FIGURE XXII–15 High auricular septal defect Adult

Note the dye passing from the superior vena cava into both auricles

more pronounced in the presence of anomalies of the pulmonary venous return than in gross defects in the auricular septum, because the primary shunt is from right to left The delineation of the aorta is of diagnostic value as it demonstrates the size of the aorta In Figure XXII–17 the aorta is clearly abnormally small

In the rare instances in which one or more of the right pulmonary veins pierce the diaphragm to enter the inferior vena cava, angiocardiography may reveal a circular shadow which extends from the lungs below the diaphragm[10] (see Figure XXII–23)

DIAGNOSIS

The diagnosis is based upon the finding of great right sided cardiac enlargement in a patient with no cyanosis and variable murmurs

The diagnosis of *a single pulmonary vein to the right auricle* is usually made only by cardiac catheterization The passage of the catheter directly into the pulmonary vein before it enters the heart establishes the anomalous entrance of

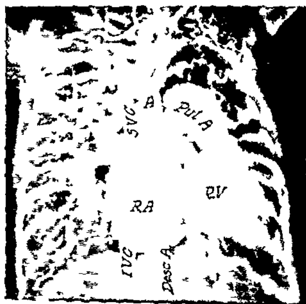


Teleoroentgenogram



Initial angiocardiogram

FIGURE XXII–16 Anomalous drainage of all the pulmonary veins into the superior vena cava Infant



Dextrogram



Lateral angiocardiogram

FIGURE XXII 1 Anomalous drainage of all the pulmonary veins into the superior vena cava (same series as Figure XXII 16) Infant

a pulmonary vein into the superior vena cava When a single pulmonary vein enters the right auricle, it will cause an increase in the oxygen content of the blood in the right auricle similar to that found in a small auricular septal defect The condition is usually asymptomatic and need cause no concern It is of clinical importance only in the differentiation of this anomaly from a small defect in the auricular septum Neither of the two conditions, however, causes symptoms

A partial anomaly of the pulmonary venous return is to be suspected when a child with a frail build and no visible cyanosis has a systolic murmur over the precordium and shows evidence of great right sided cardiac enlargement and increased pulmonary vascularity together with electrocardiographic evidence of right ventricular hypertrophy The probability of this diagnosis is further increased if, in addition, there is evidence of progressive cardiac difficulty in the absence of any history or signs of acute rheumatic fever The diagnosis is substantiated upon cardiac catheterization by the finding of a marked increase in the oxygen content of the blood in the right auricle to almost full saturation The diagnosis is also substantiated by angiocardiography if there is a filling defect along the margin of the right auricle or if dye passes simultaneously into both auricles

A partial anomaly of the pulmonary venous return with an intact auricular septum is to be suspected when the above mentioned findings of a partial anomaly of the pulmonary venous return are combined with a systolic murmur at the apex and electrocardiographic evidence not only of right ventricular hypertrophy in V_1 but of left as well as right ventricular hypertrophy in V_6

When all the pulmonary veins drain into the superior vena cava, the diagnosis can usually be made by the clinical findings combined with a characteristic "snowman seen in the x ray or upon fluoroscopy The diagnosis is readily confirmed by cardiac catheterization which shows a marked increase in the oxygen content of the sample taken from the superior vena cava in comparison with that taken from the brachial vein, or by angiocardiography which readily shows the nature of the wide mediastinal shadow

DIFFERENTIAL DIAGNOSIS

The condition may call for differentiation from pneumonia in early infancy and from other conditions which cause great right sided cardiac enlargement and minimal cyanosis, such as an auricular septal defect, a large ventricular septal defect, and a persistent ostium atrioventriculare commune, and occasionally from pure' pulmonary stenosis

Pneumonia, in early infancy, may be difficult to differentiate clinically be-

...ment causes dullness over the entire left chest and,

[illegible]

The cyanosis may not be more intense than is consistent with ... oscopy or x ray usually reveals that the dullness is cardiac, not pulmonary, in origin

An auricular septal defect does not usually cause such serious cardiac embarrassment in early infancy, and when it does the condition is generally associated with murmurs and thrills The time of onset of the cardiac enlargement differs in the two malformations If an auricular septal defect causes enlargement, the rapid increase in the size of the heart occurs shortly after birth, thereafter, until the development of a superimposed infection, no further cardiac enlargement occurs In the malformation under discussion there is little enlargement of the heart during the first months of life, thereafter the enlargement is progressive If the foramen ovale tends to seal off in the normal manner, the period of most rapid enlargement occurs between the second and sixth months of life

When, as is commonly the case, the foramen ovale remains patent, or if there is a gross defect in the auricular septum, the contour of the heart and even the physical findings may closely resemble those of an auricular septal defect of the ostium secundum type Both malformations cause great enlargement of the right auricle and the right ventricle, an abnormal fullness of the pulmonary artery, and increased hilar shadows The vascular markings are more pronounced and more widespread in the anomalies of the venous return than in auricular septal defects The hilar dance is more conspicuous The feature which differentiates the two conditions is the abnormal dilatation of the superior vena cava at the point of entrance of the pulmonary drainage If the pulmonary veins open into the posterior wall of the right auricle, the two conditions may be closely similar In both malformations there is a communication between the two auricles but the direction of the shunt is different In an auricular septal defect the shunt is normally from left to right, whereas in the anomaly under discussion the shunt is from right to left In an auricular septal defect the oxygen saturation of the arterial blood is usually normal, whereas in a total anomaly of the pulmonary venous return there is usually slight reduction in the oxygen saturation of the arterial blood When there is a partial anomaly of the pulmonary venous return, this distinction does not hold, as under such circumstances the arterial blood is usually fully saturated

By and large, anomalies of the pulmonary venous return cause greater difficulty than do gross defects in the auricular septum Hence this anomaly should

always be suspected if, in spite of all therapy, the patient remains in chronic congestive cardiac failure, especially if the failure becomes gradually more severe

A large, high ventricular septal defect may be confused with an anomaly of the pulmonary venous return When the shunt is at the ventricular level, there is usually enlargement of the left auricle and the electrocardiogram usually shows evidence of 'combined' hypertrophy in the unipolar precordial leads Cardiac catheterization will show that the shunt is at the ventricular level

A persistent ostium atrioventriculare commune may occasionally be confused with an anomaly of the pulmonary venous return, as both may cause progressive cardiac enlargement and chronic cardiac failure Cardiac catheterization will aid in the differentiation of the two conditions if all four chambers are catheterized, it is strong presumptive evidence of a persistent ostium atrioventriculare commune

'Pure" pulmonary stenosis may occasionally call for differentiation from an anomaly of the pulmonary venous return because of the fullness of the pulmonary conus The rasping systolic murmur and the weak or absent second sound should sharply differentiate these two conditions

COMMONLY ASSOCIATED ANOMALIES

Defects in the auricular septum are so common as to constitute an integral part of the anomaly of the pulmonary venous return Indeed, when there is an auricular septal defect it should never be closed without the correction of the pulmonary venous abnormality

Pulmonary valvular stenosis may occur in combination with a total anomaly of the venous return Snellen[7] has reported such a case Although the pulmonary stenosis increases the work of the right ventricle, it lessens the pulmonary blood flow and forces more blood from the right auricle to the left auricle and thence to the systemic circulation Thereby it lessens the incapacity of the patient The pulmonary stenosis should never be relieved without the correction of the anomalous pulmonary venous drainage

TREATMENT

Surgical correction of anomalies of the pulmonary venous return is now done with remarkable success under direct vision It may be done with hypothermia or with a pump and oxygenator

These anomalies so frequently occur in combination with a defect in the auricular septum that the correction of a partial anomaly of the pulmonary ve

nous return is frequently regarded as part of the operation for closure of an auricular septal defect of the ostium secundum type (see *Chapter XXIII*, Section B)

A partial anomaly of the venous return in which the right pulmonary veins enter the right auricle close to the entrance of the superior vena cava, combined with a high auricular septal defect, is probably the easiest condition to correct It is usually possible to close the auricular septal defect in such a manner that the pulmonary veins open into the left auricle without serious obstruction to the superior vena cava The danger of the latter is greatly reduced if the left superior vena cava drains into the coronary sinus, as it sometimes does in this type of anomaly

When the auricular system is intact and some of the pulmonary veins enter the right auricle, either the anomalous veins must be transplanted into the left auricle or the auricular septum must be realigned in such a manner that all the pulmonary veins drain into the left auricle Dr Henry Bahnson has successfully performed this latter type of operation with the use of a cardiopulmonary by pass

When all the pulmonary veins drain into the superior vena cava the operation is theoretically simple, but in reality the left auricle may not be sufficiently large to handle all the blood returned from the lungs The younger the patient at the time difficulty develops, the greater is the probability that the left side of the heart is seriously underdeveloped If the patient has survived to childhood there is real hope that the left ventricle may be sufficiently large to pump a normal volume of blood to the body Under such circumstances the child may be greatly aided if the auricular septal wall is realigned, as Bahnson[11] has done, so that the left auricle is increased in size and the pulmonary veins and the superior vena cava both drain into the left auricle

Surgical correction of an anomaly of the pulmonary venous return virtually restores the circulation to normal Compensation immediately improves and the heart decreases in size (compare the x rays in Figure XXII-18 and also those in Figure XXII-7) Nevertheless, the full benefit of the operation may not be attained for a year, as it takes time for the left ventricle to hypertrophy so that it can perform the increased work required by exercise as efficiently as does the normal heart

PROGNOSIS

The prognosis varies with the extent of the anomaly of the pulmonary venous return and the structure of the auricular septum Although occasionally a



Before operation



Two months after operation

FIGURE XXII–18 Partial anomaly of the pulmonary venous return and an auricular septal defect Child

total anomaly of the pulmonary venous return may be compatible with life for many years, frequently the condition leads to progressive cardiac enlargement and cardiac failure in infancy or childhood Surgical correction greatly improves the prognosis

A partial anomaly of the pulmonary venous return combined with an intact auricular septum also leads to progressive cardiac enlargement and death in early childhood Surgical correction of this anomaly changes the prognosis from hopeless to excellent

A partial anomaly of the pulmonary venous return combined with a gross defect in the auricular septum is extraordinarily well tolerated In many instances the condition is readily compatible with life for many years Nevertheless, a partial anomaly of the pulmonary venous return far more frequently causes difficulty than does an auricular septal defect of the ostium secundum type The condition is, however, amenable to surgery Re alignment of the auricular septum, so that the pulmonary ostia open into the left auricle, restores the heart and the circulation to normal and thereby greatly improves the prognosis

SUMMARY

Anomalies of the pulmonary venous return are relatively common The most common of these anomalies is a partial anomaly of the pulmonary venous return combined with a gross defect in the auricular septum When all the pulmonary veins drain into the right auricle, they enter either by way of the superior vena cava or through the coronary sinus

A single pulmonary vein causes little strain on the circulation When two or more pulmonary veins enter the right auricle, a considerable volume of oxygenated blood is pumped through the lesser circulation A total anomaly of the pulmonary venous return means that all the blood from the lungs is returned to the right auricle and the only blood which reaches the systemic circulation is that which is shunted from the right auricle to the left auricle The malformation places a great strain on the right side of the heart and usually leads to progressive cardiac enlargement and death at an early age

A partial anomaly of the pulmonary venous return with an intact auricular septum places an ever increasing load upon the heart Cardiac failure occurs early Unless corrected by surgery the condition is seldom compatible with life for more than a few years

An auricular septal defect is compensatory It aids in the direction of oxy

genated blood to the systemic circulation and thus renders the condition compatible with life

A partial anomaly of the pulmonary venous return combined with an auricular septal defect is readily compatible with life for many years

The entrance of the pulmonary veins into the right auricle means that a large amount of oxygenated blood is directed to the lungs The larger the number of pulmonary veins which enter the right auricle, the greater is the pulmonary blood flow and the smaller is the systemic blood flow

The malformation causes a large pulmonary blood flow but a small effective flow The pulmonary pressure is usually moderately elevated, but not to the systemic level The large volume of oxygenated blood returned to the right auricle and the right ventricle causes great enlargement of the right side of the heart The left side of the heart may or may not be hypoplastic

The patient has a frail build The chest shows left sided deformity or is barrel shaped Dyspnea on exertion is common The patient fatigues easily Cyanosis is usually absent but may occur in infants, especially with the terminal collapse of the circulation

Pulmonary congestion is the rule Edema is a late manifestation

The heart is enlarged, especially on the right side When the auricular septum is intact, the condition leads to rapid cardiac enlargement Generally, however, there is a defect in the auricular septum and the enlargement occurs slowly The pulmonic second sound is accentuated The murmurs are variable, a systolic murmur over the precordium and also a mid diastolic murmur and a gallop rhythm are, however, of common occurrence An apical systolic murmur is always suggestive of an intact auricular septum Cardiac arrhythmias are relatively common Cardiac failure may occur

The x ray shows enlargement of the right auricle and of the right ventricle, a full pulmonary conus, and increased vascular markings The left auricle is usually normal in size The entrance of all the pulmonary veins into the superior vena cava causes the cardiac shadow to resemble a snowman with his head off center The entrance of all the pulmonary veins into the coronary sinus frequently causes a protrusion of the upper margin of the right auricle

The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy There may be a tall R wave as well as a deep S wave in V_6 when the auricular septum is intact

The red blood cell count is usually normal

The oxygen saturation of the arterial blood is surprisingly high It is always

fully saturated in a partial anomaly of the pulmonary venous return but in a total anomaly slight unsaturation is inevitable

Cardiac catheterization may readily confirm the diagnosis when all the pulmonary veins enter the superior vena cava and aid in the diagnosis of a partial anomaly of the pulmonary venous return by the demonstration of a marked increase in the oxygen content of the blood in the right auricle The condition is to be suspected when there is an abrupt rise in the oxygen saturation of the blood in the right auricle to 90 per cent or higher The pulmonary pressure is usually moderately elevated

Angiocardiography also confirms the diagnosis of a total anomaly of the pulmonary venous return to the superior vena cava and aids in the diagnosis of a partial anomaly by the demonstration of a filling defect along the margin of the right auricle

The diagnosis should be suspected in patients who show dyspnea on exertion but no cyanosis and who show great right sided cardiac enlargement with increased pulmonary vascularity and electrocardiographic evidence of right ventricular hypertrophy When all the pulmonary veins drain into the superior vena cava, the diagnosis can usually be made by x ray or fluoroscopy When there is a partial anomaly of the pulmonary venous return, cardiac catheterization or angiocardiography may be necessary to establish the diagnosis

The condition requires differentiation from pneumonia in early infancy, from an auricular septal defect of the ostium secundum type, from a large ventricular septal defect, and from a persistent ostium atrioventriculare commune and occasionally from pure pulmonary stenosis

The commonly associated anomaly is a defect in the auricular septum, this is always compensatory in nature Pulmonary valvular stenosis may occur It, too, may be of benefit to the patient

The condition is amenable to surgery A partial anomaly of the venous return combined with a defect in the auricular septum is virtually as easily corrected as an auricular septal defect of the ostium secundum type. A total anomaly of the pulmonary venous return may also be corrected but when such anomalies cause difficulty at an early age they are frequently associated with serious underdevelopment of the left side of the heart

Surgical correction of the defect virtually restores the heart and circulation to normal

The prognosis varies with the number of the pulmonary veins which enter the right auricle with the size of the auricular defect, and with the development

genated blood to the systemic circulation and thus renders the condition compatible with life

A partial anomaly of the pulmonary venous return combined with an auricular septal defect is readily compatible with life for many years

The entrance of the pulmonary veins into the right auricle means that a large amount of oxygenated blood is directed to the lungs The larger the number of pulmonary veins which enter the right auricle, the greater is the pulmonary blood flow and the smaller is the systemic blood flow

The malformation causes a large pulmonary blood flow but a small effective flow The pulmonary pressure is usually moderately elevated, but not to the systemic level The large volume of oxygenated blood returned to the right auricle and the right ventricle causes great enlargement of the right side of the heart The left side of the heart may or may not be hypoplastic

The patient has a frail build The chest shows left sided deformity or is barrel shaped Dyspnea on exertion is common The patient fatigues easily Cyanosis is usually absent but may occur in infants, especially with the terminal collapse of the circulation

Pulmonary congestion is the rule Edema is a late manifestation

The heart is enlarged, especially on the right side When the auricular septum is intact, the condition leads to rapid cardiac enlargement Generally, however, there is a defect in the auricular septum and the enlargement occurs slowly The pulmonic second sound is accentuated The murmurs are variable, a systolic murmur over the precordium and also a mid diastolic murmur and a gallop rhythm are, however, of common occurrence An apical systolic murmur is always suggestive of an intact auricular septum Cardiac arrhythmias are relatively common Cardiac failure may occur

The x ray shows enlargement of the right auricle and of the right ventricle, a full pulmonary conus, and increased vascular markings The left auricle is usually normal in size The entrance of all the pulmonary veins into the superior vena cava causes the cardiac shadow to resemble a snowman with his head off center The entrance of all the pulmonary veins into the coronary sinus frequently causes a protrusion of the upper margin of the right auricle

The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy There may be a tall R wave as well as a deep S wave in V_6 when the auricular septum is intact

The red blood cell count is usually normal

The oxygen saturation of the arterial blood is surprisingly high It is always

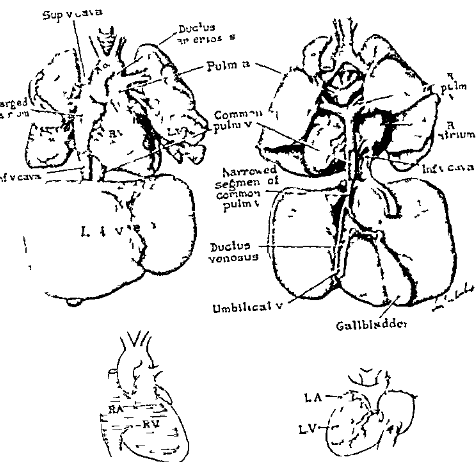


Figure xxii-19 Anomalous drainage of all the pulmonary veins into the hepatic vein Infant

Therefore the anomalous return of the pulmonary veins places little strain on the fetal circulation

The only blood which reaches the left side of the heart is that which passes from the right auricle through the foramen ovale to the left auricle It seems probable that less than the normal quota of blood enters the left auricle and the left ventricle as these chambers remain hypoplastic Nevertheless, even though less blood may be pumped out through the aorta and more blood may be pumped out through the pulmonary artery than in the normal heart, there is no great alteration in the circulation of the blood through the body of the fetus, as

of the left side of the heart Without operation, the condition may lead to slow cardiac enlargement but it is frequently compatible with life for many years Successful operation renders the prognosis excellent

B *Anomalous Drainage of the Pulmonary Veins into the Inferior Vena Cava by Way of the Hepatic Vein*

The anomalous drainage of all of the pulmonary veins into the hepatic vein occasionally occurs Under such circumstances all four pulmonary veins enter into a single trunk which pierces the diaphragm and drains into the ductus venosus or into the hepatic vein and thence into the inferior vena cava This anomaly causes a totally different clinical syndrome from that produced by the anomalous return of the pulmonary veins directly into the right auricle or into the superior vena cava Hence it is presented as a separate clinical entity

NATURE OF THE MALFORMATION

In this anomaly all four pulmonary veins unite into a single vessel which passes behind the heart, pierces the diaphragm, and enters the ductus venosus or the hepatic vein, which in turn opens into the inferior vena cava The single pulmonary vein decreases in size as it extends downward and at the point of entrance of this vessel into the ductus venosus the opening is extremely small Moreover, the ductus venosus, when it persists, is only a tiny vessel which opens into the inferior vena cava, as shown in Figure XXII–19 The small size of the opening of the pulmonary vein into the ductus venosus and the minute size of the ductus venosus render it difficult for the blood in the pulmonary veins to enter the inferior vena cava

In addition, the left auricle is extremely small Indeed, in this anomaly, as in some instances when all the pulmonary veins drain into the superior vena cava, the primary defect may be the failure of the left auricle to expand Consequently the single pulmonary vein fails to meet the outpouching of the left auricle Regardless of etiology, the left auricle is an extremely small chamber and the left ventricle is also hypoplastic Nevertheless, the great vessels arise in the normal manner from their respective ventricles and the ventricular septum is intact, the foramen ovale and the ductus arteriosus are patent at birth

COURSE OF THE CIRCULATION

During fetal life the lungs do not function and most of the blood in the pulmonary artery flows through the ductus arteriosus to the systemic circulation

ventricle become tremendously enlarged, the left auricle and the left ventricle remain hypoplastic, pulmonary congestion is marked

CLINICAL FINDINGS

Cyanosis is present at birth and persists throughout the infant's brief life

Dyspnea is also present from birth and may be severe

The liver becomes engorged As long as the heart action *remains vigorous* pulsations may be palpable at the margin of the liver

Edema occurs early and is severe

CARDIAC FINDINGS

The heart appears normal in size Although the right auricle and the right *ventricle are huge, the* left auricle and the left ventricle are so small that the cardiac shadow is not enlarged The *second sound* to the *left of the sternum* at the base of the heart is usually accentuated There may be a gallop rhythm

Murmurs are usually absent over the precordium but may be heard over the liver that is, below the diaphragm

Cardiac failure occurs early This is virtually the only malformation in which cardiac failure causes edema of the extremities in the absence of demonstrable cardiac enlargement.

X RAY AND FLUOROSCOPIC FINDINGS

The heart is remarkably normal in size and shape (see Figure XXII–20) The vascular markings are extraordinarily heavy and diffuse throughout the lungs At first glance these shadows appear more like miliary tuberculosis than like pulmonary congestion Careful study will usually show that the dense pulmonary markings are composed of central areas of congestion with lines radiating out in all directions as do the spokes of a bicycle wheel

ELECTROCARDIOGRAPHIC FINDINGS

The standard leads show a right axis deviation and the unipolar precordial leads show evidence of extreme right ventricular hypertrophy The P waves may, in addition, be high and peaked

SPECIAL TESTS

The condition of the infant is frequently too precarious to permit extensive studies

the ductus arteriosus directs a considerable volume of blood to the systemic circulation

After birth with the expansion of the lungs, blood flows freely to the lungs but difficulty is encountered in the return of the blood to the heart Not only do the pulmonary veins fail to open into the left auricle, but the small size of the ductus venosus and the small caliber of the single pulmonary vein which enters the ductus venosus severely obstruct the pulmonary venous return

All the blood which is returned from the lungs enters the inferior vena cava and thence is returned to the right auricle In addition, all the blood from the body is returned by the superior vena cava and the inferior vena cava to the right auricle Inasmuch as no blood is returned directly to the left auricle, the pressure in the left auricle remains low and some blood from the right auricle is forced through the foramen ovale to the left auricle and thence to the left ventricle and is pumped out by way of the aorta to the systemic circulation

Although the high pressure in the right side of the heart may tend to keep the ductus arteriosus open, in most instances the ductus arteriosus eventually undergoes normal obliteration

Inasmuch as all the blood from the body and from the lungs is returned to the right auricle and thence is directed to the lungs and to the systemic circulation, the oxygen content of the blood in the aorta and that in the pulmonary artery are the same Furthermore, since there is difficulty in the return of the oxygenated blood from the lungs through the inferior vena cava, only a small volume of oxygenated blood is mixed with a large volume of venous blood, cyanosis is moderately intense The course of the circulation is shown in Diagram XXII–5

PHYSIOLOGY OF THE MALFORMATION

The difficulty in the return of the venous blood from the lungs to the heart causes serious back pressure in the lungs This in turn raises the pressure in the pulmonary vascular bed and causes pulmonary hypertension Therefore, even if the ductus arteriosus undergoes normal obliteration, the pressure in the lungs remains high The right ventricle is required not only to pump an abnormally large volume of blood but also to pump against increased resistance The high pressure in the right ventricle is reflected back into the right auricle and this in turn aids in the direction of blood through the foramen ovale to the left auricle, the left ventricle, and the systemic circulation Nevertheless, the malformation places a great strain on the right side of the heart The right auricle and the right

ventricle become tremendously enlarged, the left auricle and the left ventricle remain hypoplastic, pulmonary congestion is marked

CLINICAL FINDINGS

Cyanosis is present at birth and persists throughout the infant's brief life

Dyspnea is also present from birth and may be severe

[illegible] the heart action remains vigorous

[illegible]

Edema occurs early and is severe

CARDIAC FINDINGS

The heart appears normal in size Although the right auricle and the right ventricle are huge, the left auricle and the left ventricle are so small that the cardiac shadow is not enlarged The *second sound* to the left of the sternum at the base of the heart is usually accentuated There may be a gallop rhythm

Murmurs are usually absent over the precordium but may be *heard over the* liver, that is, below the diaphragm

Cardiac failure occurs early This is virtually the only malformation in which cardiac failure causes edema of the extremities in the absence of demonstrable cardiac enlargement

X RAY AND FLUOROSCOPIC FINDINGS

The heart is remarkably normal in size and shape (see Figure XXII-20) The vascular markings are extraordinarily heavy and diffuse throughout the lungs At first glance these shadows appear more like miliary tuberculosis than like pulmonary congestion Careful study will usually show that the dense pulmonary markings are composed of central areas of congestion with lines radiating out in all directions as do the spokes of a bicycle wheel

ELECTROCARDIOGRAPHIC FINDINGS

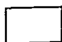
The standard leads show a right axis deviation and the unipolar precordial leads show evidence of extreme right ventricular hypertrophy The P waves may, in addition, be high and peaked

SPECIAL TESTS

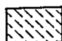
The condition of the infant is frequently too precarious to permit extensive studies

Diagram XXII–5

Head and upper extremities
R Lung
P.A.
S.V.C.
P.V.
Aorta
Pul A.
L Lung
Pul V
Pulmonary Artery
Foramen Ovale
L.A.
R.A.
Pulmonary Vein
L.V.
Descending Aorta
Liver
R.V.
Ductus Venosus
I.V.C.
Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

DIAGRAM XXII-5

Anomalous drainage of all the pulmonary veins into the right auricle by way of the hepatic vein

The essential feature of this anomaly is that all the pulmonary veins join together and pierce the diaphragm as a single pulmonary vein which enters the hepatic vein or the ductus venosus, which in turn opens into the inferior vena cava Furthermore, the pulmonary vein becomes gradually reduced in size, so that there is but a small opening into the hepatic vein

Most of the blood from the right auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs All the blood from the lungs is returned by a single pulmonary vein to the right side of the heart by way of the hepatic vein and the inferior vena cava Inasmuch as no blood is returned to the left auricle in the normal manner and all the blood is returned to the right auricle, the pressure in the right auricle is greater than that in the left auricle and some blood flows through the foramen ovale to the left auricle and thence to the left ventricle. The blood in the left ventricle is pumped out through the aorta to the systemic circulation and is returned by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again The right auricle and the right ventricle become enlarged and the left auricle and the left ventricle remain small The increased volume of blood which is pumped out to the lungs encounters great difficulty in its return through the hepatic vein The pressure in the pulmonary veins rises and this in turn increases the resistance in the lungs which still further increases the work of the right side of the heart The lungs fail to expand normally and the blood which is returned from the lungs may not be fully saturated

Clinical diagnosis The infant is cyanotic and dyspneic Cardiac failure with congestion in the lungs engorgement of the liver and edema of the extremities occur before there is demonstrable cardiac enlargement. The x ray picture resembles that of miliary tuberculosis but the infant is in severe cardiac failure Death usually occurs between seventeen and twenty two days of age



FIGURE XXII–20 Anomalous drainage of all the pulmonary veins into the hepatic vein (same patient as in Figure XXII–21) Infant

Angiocardiography reveals prompt filling of the right side of the heart and of the pulmonary artery It may also reveal a conglomeration of dye below the diaphragm as it accumulates at the point of constriction where the single vein enters the hepatic vein (see Figure XXII–21)

DIAGNOSIS

The diagnosis is based upon the finding of dyspnea and cyanosis and early evidence of cardiac failure and edema of the extremities in an infant with a heart of normal size Usually no murmurs are audible The diagnosis is substantiated by x ray evidence of diffuse congestion in the lungs and electrocardiographic evidence of extreme right ventricular hypertrophy

DIFFERENTIAL DIAGNOSIS

Complete transposition of the great vessels may be considered because of the early signs of dyspnea, cyanosis, and cardiac failure The mistaken diagnosis may appear to be substantiated by the x ray findings of heavy vascular markings which extend to the periphery of the lungs A complete transposition of the great vessels, however, seldom if ever causes difficulty until the heart is greatly enlarged Therefore, when the heart is normal in size and the infant develops car



FIGURE XXII–21 Anomalous drainage of all the pulmonary veins into the hepatic vein (same patient as in Figure XXII–20) Infant

The arrow points to the accumulation of dye below the diaphragm

diac failure, an anomalous return of the pulmonary veins to the hepatic vein is the most probable diagnosis

Miliary tuberculosis may be considered because of the diffuse, fluffy shadows in the lungs and the normal size of the heart. The early onset of edema indicates that the difficulty is cardiac in origin

TREATMENT

Surgical correction is extremely difficult. The author has been told of one infant in whom at fifteen days of age the anomalous pulmonary veins from the left lung were anastomosed to the left auricle,[12] for the ensuing two months he did well and then died of an aspiration pneumonia

In many instances the small size of the left auricle and the hypoplasia of the left ventricle pose a serious obstacle to the correction of the malformation Under such circumstances, it is possible that a direct anastomosis of the pulmonary vein to the *posterior wall of the right auricle* would lessen the congestion in the lungs and aid in the return of oxygenated blood to the heart

PROGNOSIS

The prognosis is poor Most infants with this anomaly die between the seventeenth and the twenty-second day of life The author has known one infant who lived for three months but such cases are extremely rare

SUMMARY

The anomalous return of all the pulmonary veins into the inferior vena cava is always associated with obstruction of the pulmonary venous return as well as with underdevelopment of the left side of the heart Pulmonary congestion is marked and pulmonary pressure remains high

Cyanosis and dyspnea date from birth Engorgement of the liver and edema of the extremities occur early The heart remains normal in size Pulmonary congestion is so severe and widespread as to suggest miliary tuberculosis

Cardiac failure occurs early and before cardiac enlargement is demonstrable Surgical correction is extremely difficult Most infants die at approximately three weeks of age

C *The Scimitar Syndrome*

The anomalous drainage of some or all of the right pulmonary veins into the inferior vena cava produces an entirely different clinical syndrome when it occurs in combination with defective development of the right lung than when the anomalous pulmonary venous return is the primary malformation The clinical syndrome of hypoplasia of the right lung associated with the anomalous development of the right pulmonary artery and veins accompanied by displacement of the heart was first described by Park in 1912 [13] Recently a number of cases have been reported McKusick and Cooley[10] and Steinberg[14] have reported the characteristic radiological findings and Halasz et al [15] have reviewed the literature and added three cases For a detailed discussion of this syndrome the reader is referred to the above reports and to the report by Neill, Ferencz, Sabiston, and Sheldon [16]

NATURE OF THE MALFORMATION

The malformation concerns the development of the right lung, its arterial blood supply, and its venous return Part of the right lung is grossly underdeveloped it fails to establish its normal connection with the pulmonary artery and retains its embryonic blood supply through the bronchial arteries The abnormality also affects the venous return Although one or more of the pulmonary

veins may empty into the left auricle in the normal manner, the majority of the pulmonary veins unite into a common vessel which swings downward and pierces the diaphragm and enters the inferior vena cava As in all malformations, there are all grades of severity The abnormality may be so slight that the condition is inconsequential and causes no symptoms throughout life On the other hand, the hypoplasia of the right lung may be so severe that virtually the entire oxygenation of the blood takes place in the left lung and the heart is displaced into the right thoracic cavity It is this latter condition with which this section is concerned

When the right lung is grossly hypoplastic, its arterial and bronchial blood supply and its venous return are grossly abnormal The right pulmonary artery is abnormally small and directs venous blood to a small portion of the lung, usually to the upper lobe and sometimes (as in Case XXII-2) to the upper lobe and part of the middle lobe The remainder of this right lung is supplied with arterial blood by anomalous arteries which arise from the descending aorta above or below the diaphragm Furthermore, although occasionally some oxygenated blood may be returned in the normal manner to the left auricle, usually the pulmonary veins from all three lobes of the right lung unite in a single vessel which passes downward and pierces the diaphragm and empties into the inferior vena cava as shown in Figure XXII-22

Whenever the right lung is small and poorly developed, the heart is displaced into the right thoracic cavity and the normal left lung is overexpanded

COURSE OF THE CIRCULATION

The blood from the right auricle flows into the right ventricle and thence is pumped out into the pulmonary artery The blood in the left pulmonary artery flows to the left lung, where it is oxygenated, and returned in the normal manner to the left auricle, thence it flows to the left ventricle and is pumped out through the aorta to the systemic circulation and returned by the superior vena cava and the inferior vena cava to the right auricle Inasmuch as the right pulmonary artery is small and usually goes to the upper lobe only, the volume of blood which reaches the right lung is greatly reduced The middle and lower lobes are supplied by an anomalous vessel which arises from the descending aorta and carries arterial blood to those lobes Usually the veins from all three lobes of the right lung join together in an anomalous channel which pierces the diaphragm and enters the inferior vena cava Thereby some oxygenated blood from the right lung is directed into the inferior vena cava and is returned to the right

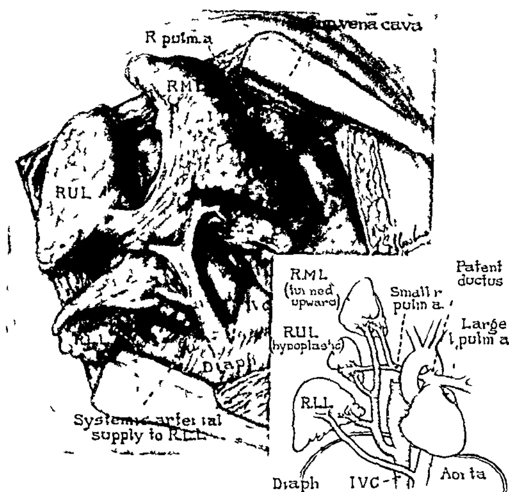


FIGURE XXII–22 Anomalous arterial blood supply to a hypoplastic right lung and anomalous drainage of the right pulmonary veins into the inferior vena cava Scimitar syndrome, drawing made at operation (Case XXII–2)

auricle There the cycle starts again The course of the circulation is shown in Diagram XXII–6

PHYSIOLOGY OF THE MALFORMATION

The physiology of the malformation concerns the abnormality of the lung even more than that of the heart Only a small portion of the right lung receives venous blood from the pulmonary artery and consequently that is the only portion which is of functional importance in the oxygenation of the blood The remainder of the right lung receives fully oxygenated blood and therefore is not able to take up oxygen or give off carbon dioxide Furthermore, almost the entire

circulation to the right lung is returned by an anomalous channel to the inferior vena cava and thence *to* the right auricle Inasmuch as the circulation *to* the right lung is markedly reduced, the volume of oxygenated blood returned to the right auricle is small Consequently the malformation places as little strain on the heart as does the entrance of a single pulmonary vein into the right auricle

It is, however, important to appreciate that the anomaly is a vascular one, although the bronchi may branch abnormally, they lead to the alveoli in the normal manner and consequently both lungs play a role in ventilation Nevertheless, the left lung, which is overexpanded and often herniates into the right thoracic cavity, is the lung of physiological importance in the exchange of oxygen and carbon dioxide

The heart, too, is displaced to the right, it is, however, normally formed

CLINICAL FINDINGS

The patient frequently suffers from repeated respiratory infections in infancy and early childhood and may suffer from bronchitis or bronchiectasis Adults are usually asymptomatic

Left sided chest deformity may be secondary to the overexpansion of the left lung

The abdominal organs occupy their normal positions

CARDIAC FINDINGS

The heart is usually displaced into the right chest, but unless complicated by some additional anomaly, the cardiac findings are basically normal The *apex* of the heart is palpable to the right of the sternum The *sounds* are of good quality The *pulmonic second sound* is normal No thrill is palpable No murmurs are audible

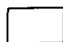
X RAY AND FLUOROSCOPIC FINDINGS

The x ray findings are so characteristic that the diagnosis can often be made from a single x ray film [10] The heart is displaced to the right and beyond its outer right margin a scimitar shadow is usually visible, extending down to the diaphragm (see Figure xxii–23) Occasionally, when there is extensive inflammatory reaction in the right lung, this shadow is not visible The left lung is over expanded and partially herniated into the right pleural cavity

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram is normal

Diagram xxii–6


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

DIAGRAM XXII–6

Scimitar syndrome

In this malformation the anomalous pulmonary venous drainage is associated with a malformation of the right lung The right lung is hypoplastic and both its arterial blood supply and its venous return are grossly abnormal The latter is by way of an anomalous channel which pierces the diaphragm and enters the inferior vena cava. The heart which is displaced to the right, is normally formed, the left lung is over expanded and frequently is herniated into the right hemithorax

The blood from the right auricle flows into the right ventricle and is pumped out into the pulmonary artery The blood flows to the left lung in the normal manner but since the right pulmonary artery is abnormally small and the right lung is hypoplastic, only a small volume of venous blood flows to the right lung The blood which goes to the left lung is oxygenated and returned in the normal manner to the left auricle. Thence it flows to the left ventricle and is pumped out through the aorta to the systemic circulation and returned in the normal manner to the right auricle.

Inasmuch as part of the right lung is supplied by an arterial vessel which arises from the descending aorta some blood from the systemic circulation is directed to the right lung This fully oxygenated blood circulates through the right lung but is of no functional importance All the veins from the right lung unite in a common venous channel which pierces the diaphragm and enters the inferior vena cava The veins which drain the right lung direct fully oxygenated blood into the inferior vena cava and this blood together with the venous blood returned by the inferior vena cava, enters the right auricle There the cycle starts again Inasmuch as a relatively small amount of blood is directed to the right lung, only a small amount of oxygenated blood is returned by way of the inferior vena cava to the right auricle. The malformation places virtually no strain upon the circulation

Clinical diagnosis The condition is to be suspected in an asymptomatic patient in whom the heart is displaced to the right and the left lung is overexpanded The diagnosis can be confirmed by a single x ray film if the heart is seen to be displaced to the right and the scimitar shadow is seen to extend to the diaphragm beyond the right margin of the cardiac silhouette

When such a shadow is not present angiocardiography may be necessary to demonstrate the anomalous venous drainage into the inferior vena cava

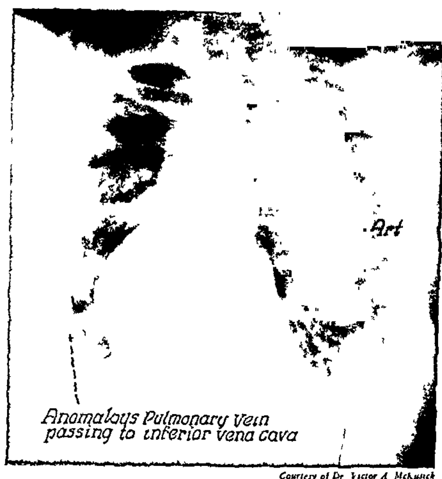


FIGURE XXII-23 Scimitar shadow caused by anomalous drainage of the right pulmonary veins into the inferior vena cava (teleoroentgenogram) Adult

SPECIAL TESTS

Additional studies may be necessary to establish the diagnosis if the "scimitar" shadow is not visible in the x ray

The measurement of the *vital capacity* is of no aid, it is normal

Cardiac catheterization shows that the heart is displaced to the right but the catheter enters the right auricle, the right ventricle, and the pulmonary artery in the normal manner If repeated samples of blood are taken from various levels in the inferior vena cava, at the point where the anomalous vein drains into the inferior vena cava, fully oxygenated blood is obtained

Angiocardiography[10, 14] reveals that the left pulmonary artery is markedly enlarged and the right pulmonary artery, if delineated, is exceptionally small The films taken as the dye is returned to the left side of the heart show the opaci

fication of the abnormal vessel passing from the right lung to the inferior vena cava (see Figure XXII–24)

DIAGNOSIS

The diagnosis is based upon the finding of extreme dextroposition of the heart with overexpansion of the left lung and the characteristic scimitar shadow to the right of the heart which curves downward to the diaphragm

DIFFERENTIAL DIAGNOSIS

The condition requires differentiation from true dextrocardia and dextrorotation, from situs inversus, from other types of displacement of the heart, and from agenesis of the right lung

Dextrocardia shows a characteristic electrocardiographic pattern the P waves and the ventricular complexes in Lead I are inverted and Leads II and III replace each other In this syndrome the upright P waves in Lead I clearly indicate that the condition is not a dextrocardia, the heart is only displaced

Dextrorotation causes displacement of the apical thrust toward the outer right border of the cardiac dullness The liver occupies its normal position

Situs inversus means that not only the heart but also the abdominal organs are transposed

Other types of displacement of the heart usually show some pulmonary cause for the displacement and do not reveal the scimitar shadow

Agenesis of the right lung causes displacement of the heart and the overexpansion of the left lung obliterates the cardiac shadow (see Figure XXII–25), furthermore, there is no scimitar shadow

TREATMENT

Prophylactic chemotherapy or antimicrobial agents are helpful in the early months of life, when frequent and severe respiratory infections are a menace The malformation itself cannot be corrected Lobectomy or pneumonectomy may be indicated if there is evidence of bronchiectasis

PROGNOSIS

The prognosis is determined by the pulmonary pathology rather than by the cardiac abnormality Most patients outgrow their susceptibility to respiratory infections, thereafter the prognosis is excellent The basic anomalies of this syndrome are compatible with a long and active life



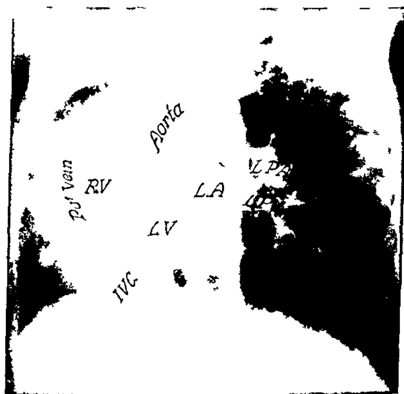


FIGURE XXII-24 Scimitar shadow opacified by angiocardiography Anomalous drainage of the right pulmonary veins into the inferior vena cava (Case XXII-2) Child

SUMMARY

The anomalous drainage of the right pulmonary veins into the inferior vena cava occurs in association with a gross abnormality in the development of the right lung, displacement of the heart into the right thoracic cavity, and overexpansion of the left lung

The malformation is primarily one which concerns the development of the right lung The right lung is hypoplastic and only a small part is of functional importance in the oxygenation of the blood In addition, most if not all the blood from the hypoplastic lung is returned by an anomalous vessel which pierces the diaphragm and enters the inferior vena cava The blood so shunted is returned to the right auricle

The heart, although displaced into the right hemithorax, is normally formed The abnormal pulmonary venous channel carries so little blood that the malformation places no detectable strain upon the heart

The left lung is overexpanded and frequently herniated into the right thoracic cavity

Infants with this abnormality frequently suffer from repeated respiratory infections

The heart is displaced to the right but is otherwise normal

X ray and fluoroscopy show the displacement and the overexpansion of the left lung and usually show the characteristic scimitar shadow to the right of the heart

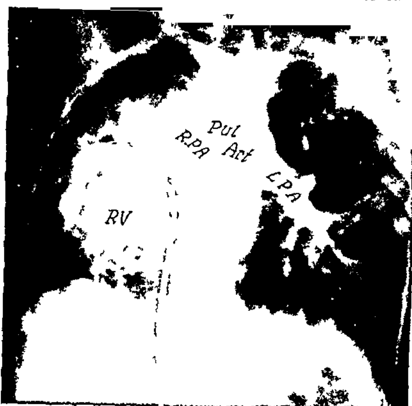
Cardiac catheterization shows a point at which fully oxygenated blood is obtained from the inferior vena cava

Angiocardiography reveals a small right pulmonary artery and the characteristic "scimitar shadow

The condition calls for differentiation from dextrocardia and dextrorotation from situs inversus, from other types of displacement of the heart, and from other pulmonary abnormalities, especially agenesis of the right lung

The diagnosis is based on the x ray findings of displacement of the heart into the right thoracic cavity and the characteristic scimitar shadow beyond the right cardiac border which curves toward the diaphragm When this is obscured by pulmonary pathology, the diagnosis can be made by angiocardiography or cardiac catheterization

Treatment is directed toward the prevention of respiratory infections The heart is basically normal, the small amount of oxygenated blood returned to the right side of the heart causes no difficulty and requires no treatment



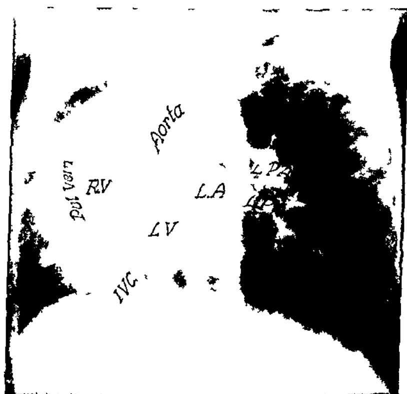


FIGURE XXII–24 'Scimitar' shadow opacified by angiocardiography Anomalous drainage of the right pulmonary veins into the inferior vena cava (Case XXII–2) Child

Prognosis, unless complicated by pulmonary pathology or some additional malformation of the heart, is excellent

Illustrative Cases

The following cases have been reported in detail by Neill, Ferencz, Sabiston, and Sheldon[18]

Case XXII-1 H T White male (age thirty-one years) The father of Case XXII-2.

History The only significant feature was that his mother advised us that as a child he had had many severe respiratory infections and had had pneumonia twice within the first five years of life He had been refused for military service because his heart was in the right side of his chest He was completely asymptomatic and as an adult had enjoyed good health

Physical examination The patient was a thin well-developed man with no dyspnea and no chest deformity The cardiac pulsations were felt to the right of the sternum but the apex beat was palpable about 2 cm to the left of the mid sternal line The heart sounds were louder to the right than to the left of the sternum The second sound over the pulmonary area showed a normal degree of reduplication, it was not accentuated

The x ray confirmed the clinical impression that the heart was displaced into the right chest but that the apex lay to the left of the mid line (see Figure XXII-6) The pulmonary vascularity was normal The aorta arched to the left

Angiocardiography showed a "scimitar shaped" shadow in the right chest such as was reported by McKusick and Cooley[10] as *characteristic* of the right pulmonary veins as they coalesced to form a common trunk which pierced the diaphragm and entered the inferior vena cava

Bronchography performed by Dr D Torrance, showed that the bronchi to the right lung were anomalous There was one large anterior bronchus which supplied the middle and upper lobes It was impossible to determine whether the upper or the middle lobe was absent but the two major divisions of the single large bronchus suggested that the upper lobe was missing

Cardiac catheterization revealed a localized area in the inferior vena cava below the diaphragm where the blood samples showed an oxygen saturation of 95 per cent The pressure in the right ventricle and that in the pulmonary artery were normal

Diagnosis Congenital displacement of the heart combined with an anomaly of the right bronchi and anomalous return of the right pulmonary veins to the inferior vena cava.

Disposition In view of the patient's excellent clinical condition no treatment was indicated or considered advisable

Comment This case illustrates the classic findings of this syndrome uncomplicated by other conditions it also illustrates its benign course.



Anterior posterior position



Right lateral position

Figure XXII–25 Congenital absence of the right lung and overexpansion of the left lung Infant

Note the absence of the cardiac shadow

Prognosis, unless complicated by pulmonary pathology or some additional malformation of the heart, is excellent

Illustrative Cases

The following cases have been reported in detail by Neill, Ferencz, Sabiston, and Sheldon [16]

CASE XXII-1 H T White male (age thirty-one years) The father of Case XXII-2

History The only significant feature was that his mother advised us that as a child he had had many severe respiratory infections and had had pneumonia twice within the first five years of life He had been refused for military service because his heart was in the right side of his chest He was completely asymptomatic and as an adult had enjoyed good health

Physical examination The patient was a thin well-developed man with no dyspnea and no chest deformity The cardiac pulsations were felt to the right of the sternum but the apex beat was palpable about 2 cm to the left of the mid sternal line. The heart sounds were louder to the right than to the left of the sternum The second sound over the pulmonary area showed a normal degree of reduplication it was not accentuated

The x ray confirmed the clinical impression that the heart was displaced into the right chest but that the apex lay to the left of the mid line (see Figure XXII-6) The pulmonary vascularity was normal The aorta arched to the left

Angiocardiography showed a scimitar shaped shadow in the right chest such as was reported by McKusick and Cooley[10] as characteristic of the right pulmonary veins as they coalesced to form a common trunk which pierced the diaphragm and entered the inferior vena cava

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Comment This case illustrates the classic findings of this syndrome uncomplicated by other conditions it also illustrates its benign course



Child

Adult
(father of child)



FIGURE XXII-26 Anomalous drainage of the right pulmonary veins into the inferior vena cava (Cases XXII-1, 2) Father and daughter

CASE XXII-2 S T (Harriet Lane Home, No B-30991) White female The daughter of Case XXII-1 Born in October, 1955 and first seen in the Cardiac Clinic in June, 1956, at eight months of age.

Chief complaint Dextrocardia with a cardiac murmur and severe respiratory infection

History The father was known to have a dextrocardia The infant had suffered from severe respiratory infection

Physical examination (at eight months of age) Weight 7 2 kg blood pressure 95/40 mm of mercury

The infant was moderately well nourished The color was normal

The positive physical findings were related to the chest

The heart was markedly enlarged and occupied most of the right chest The apex beat could not be localized and no thrill was palpable A systolic murmur was audible over the entire right chest, front and back, it was of maximal intensity to the right of the sternum The systolic murmur was thought to be continuous with an early diastolic murmur The second sound at the base of the heart to the right of the sternum was markedly accentuated but was not reduplicated The lungs were filled with rales these were more numerous over the right lung than the left but no definite area of consolidation was detected The liver was palpable 1 cm below the right costal margin The pulses in the arm and the leg were full and equal

The x ray showed that the heart filled most of the right chest cavity (see Figur XXII-26) The cardiothoracic ratio was 58 per cent The left pulmonary artery appeare large and the right pulmonary artery could not be identified The pulmonary vascu larity appeared to be within normal limits Barium swallow showed a left aortic arc and slight left auricular enlargement

The electrocardiogram showed a right axis deviation and evidence of marked rig ventricular hypertrophy The P waves were inverted in Lead 1 and upright in t augmented right arm lead

Cardiac catheterization in February 1958 showed that the heart was displaced the right but that the right auricle and the right ventricle were in normal relation each other The pressures in the pulmonary artery and in the right ventricle w markedly elevated The maximum difference in the oxygen content of the blood s ples taken from the right side of the heart was only 1 volume per cent The pres in the right ventricle was 85/6 mm of mercury and that in the pulmonary artery 85/50 mm of mercury Thus cardiac catheterization confirmed the clinical diag of pulmonary hypertension

Angiocardiography performed by Dr G Schultz in February 1958, revealec basic nature of the abnormality [10, 14] Angiocardiography was performed by the i tion of dye directly into the right ventricle. This confirmed the displacement o heart to the right The main pulmonary artery and the left pulmonary artery

markedly enlarged, the right pulmonary artery was approximately half the size of the left The right pulmonary veins were seen to form a trunk which lay behind the heart and drained below the diaphragm, forming the characteristic scimitar shadow (see Figure xxii-24) The left pulmonary veins entered the left auricle normally, the left ventricle and the aorta appeared normal

Clinical impression Dextrocardia with pulmonary hypertension and patency of the ductus arteriosus and probably some additional anomaly, complicated by bronchopneumonia

Course The infant recovered satisfactorily from her pulmonary infection with appropriate antibiotic therapy but during the ensuing year she suffered repeatedly from severe pulmonary infections

The cardiac findings remained essentially unchanged except that the continuous murmur which was audible at the first examination was thereafter inconstantly heard

Because of the repeated pulmonary infections and the probability that the right lung was hypoplastic, bronchography was performed in January, 1958, by Dr D Torrance The right main bronchus branched in an anomalous manner A single large branch apparently supplied the superior and inferior portions of the right lung anteriorly and a small bronchus which corresponded to the normal bronchus to the right upper lobe, supplied only a small area of that lobe The right lung, especially the right upper lobe, appeared markedly hypoplastic

The combination of these studies clearly showed that the child had hypoplasia of the right lung with anomalous pulmonary venous drainage and pulmonary hypertension

The occurrence of repeated severe pulmonary infections suggested the incipient development of bronchiectasis In addition, the question presented itself whether the abnormality in the right lung might be responsible for the pulmonary hypertension Therefore pneumonectomy was recommended

The operation was performed by Dr D Sabiston The right lung was found to be hypoplastic Careful dissection revealed one large artery and several small arteries which entered the lung through the diaphragm and therefore were thought to arise from the descending aorta, in addition, a systemic artery was found which extended from the upper mediastinum to the lungs The entire pulmonary venous drainage from the right lung entered a single large pulmonary vein which passed through the diaphragm near the inferior vena cava Figure xxii-22 shows the right lung and its relation to the heart as seen at operation All these vessels were individually ligated and divided and a pneumonectomy was performed The child died a few hours after operation

Autopsy No 28053 A partial autopsy performed by Dr H Sheldon showed a shift of the heart and the mediastinum to the right The surgical removal of the right lung had left a smooth pleural cavity in which the only abnormalities were the vessels which

perforated the diaphragm The largest of these vessels could be demonstrated to be a vein which entered the inferior vena cava The smaller vessels were found to arise from the abdominal aorta above the celiac axis

Examination of the heart showed that the superior vena cava and the inferior vena cava entered normally into the right auricle The foramen ovale was sealed The tricuspid valve was normal The right ventricle was dilated and hypertrophied Its wall measured 1 cm in thickness The right ventricular outflow tract was hypertrophied The pulmonary valves were normal The pulmonary artery was greatly dilated The right branch of the pulmonary artery was considerably smaller than the left The left pulmonary veins entered the left auricle in the normal manner The left ventricle was slightly hypertrophied and the aortic valve was bicuspid The aorta was normal The ductus arteriosus which was only 2 mm in diameter was patent

Examination of the left lung showed greatly dilated pulmonary vessels which branched into both lobes in the normal manner

Examination of the right lung showed three lobes Perfusion of the afferent and efferent vessels showed that the small right pulmonary artery supplied the small upper lobe and also the middle lobe. Injection of the largest of the anomalous vessels which entered the lung through the diaphragm showed that it supplied the lower lobe, which was the largest of the three. The pulmonary veins from all three lobes united to form a common trunk which extended downward and pierced the diaphragm and entered the inferior vena cava

There was also a congenital malformation of the urinary tract on the left a complete double ureter and on the right two ureters from the upper and lower poles joined together a short distance below the pelvis of the lobulated kidney

Microscopic examination of the lungs showed that the upper lobe of the right lung was essentially normal The middle and lower lobes, which had an aberrant blood supply, showed some pathology In contrast to these relatively benign findings the left lung showed evidence of severe pulmonary arteriolar disease

Comment Autopsy confirmed that the basic abnormality was hypoplasia of the right lung associated with anomalous pulmonary drainage from that lung to the inferior vena cava combined with displacement of the heart into the right thoracic cavity and overexpansion of the left lung

In addition, it appeared that the severe pulmonary vascular disease of the *left* lung caused the severe pulmonary hypertension The latter undoubtedly contributed to the severity of the pulmonary infection to the adverse clinical course and to the death of the child

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CHAPTER XXIII

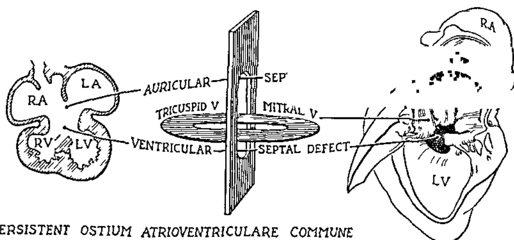
AURICULAR SEPTAL DEFECTS

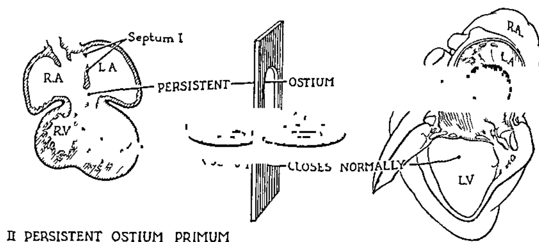
THERE are various types of defects which involve the auricular septum The abnormality may be great or slight There may be but a single auricle or the auricular septum may be normally formed and the foramen ovale held open by the high pressure on the right side of the heart Such patency of the foramen ovale should, however, be distinguished from an auricular septal defect A defect high up in the auricular septum, which is commonly spoken of as a defect of the ostium secundum type, should be differentiated from one which lies at the base of the auricular septum, that is, a persistent ostium primum' The latter type of defect merges into a persistent ostium atrioventriculare commune The last two defects are frequently classed as endocardial cushion defects or as defects of the A V canal

EMBRYOLOGY

A brief review of the development of the auricular septum will aid in the differentiation of these various conditions and will clarify the terminology

When the primitive cardiac loop first forms, there is but a single auricle and a single ventricle When an arrest in the development of the atria occurs at this stage, there is but a single auricle (see Section A) Normally the auricular septum grows down from above and the ventricular septum grows up from the base of the ventricle, the atrioventricular canal leads from the auricle to the ventricle On either side of this canal the endocardial cushions destined to form the mitral and the tricuspid valves make their appearance An arrest in the development of the heart at this stage results in a *persistent ostium atrioventriculare commune*, a defect which involves both the auricular and ventricular septa (see Figure XXIII–1) The mitral and tricuspid valves either fuse together through the defect or they remain undifferentiated, forming a common atrioventricular valve (see Figure XXIII–2) When the valves are closed, there is a defect in the ventricular septum and also one in the auricular septum When the valves are open, there is free communication between all four chambers Clinically this malformation more closely resembles a defect in the ventricular septum than one high up in the auricular septum In all other types of auricular septal defects, the ventricular septum is intact





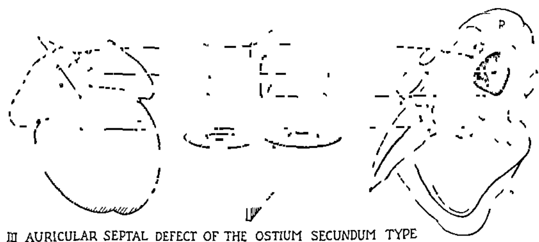


Figure xxiii-1 Defects in the auricular septum Embryological drawing after Patten[2]

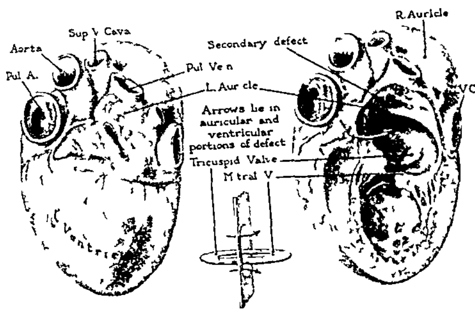


Figure XXIII 2 Persistent ostium atrioventriculare commune Infant

A persistent ostium primum is a defect which is caused by the failure of the first primitive opening in the auricular septum to close This opening lies at the base of the auricles, just above the mitral and tricuspid valves (see Figure XXIII-1) Indeed, this malformation represents a slightly later arrest in the development of the auricular septum than does an ostium atrioventriculare commune Not infrequently the arrest occurs before the mitral and tricuspid valves are completely formed Hence cleavage of one of these valves, especially that of the mitral valve, is extremely common in this malformation (see Figure XXIII-3) Furthermore, the base of the defect is formed by these valves (see Figure XXIII-4) If the mitral and tricuspid valves are normal, the clinical syndrome is similar to that of an auricular septal defect which occurs higher up in the auricular wall If the mitral valve is insufficient, the malformation is distinctive

Normally, as the embryo grows, the ostium primum closes and the ostium secundum opens It is the ostium secundum which persists as the foramen ovale, the valve which covers it is formed on the left side of the auricular septum Failure of the valve to become completely sealed after birth results in a condition which is totally different from that which occurs when there is a defect in the formation of the auricular septum during embryonic life

A defect in the auricular septum is due to an abnormality in the formation of the septum In contrast to a patency of the foramen ovale, there is a gross defect

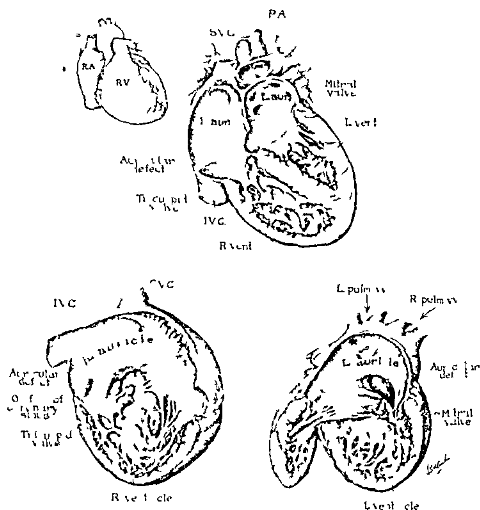


FIGURE XXIII-3 Persistent ostium primum with clefts in the mitral and tricuspid valves (same patient as in Figures XXIII-17, 18) Child

in the auricular wall (see Figure XXIII-1) The opening is not covered by a valve or membrane Consequently there is free communication between the two auricles Such a defect permits the flow of blood in either direction, but, inasmuch as after birth the pressure in the left auricle is normally slightly greater than that in the right auricle, the shunt is usually from left to right In contrast to this, in cases in which the valve covering the foramen ovale is not completely sealed, the shunt can only be from right to left

The defect in the auricular septum frequently occupies a position close to the foramen ovale, in some instances it may be similar to the foramen ovale in size and shape Even though the defect superficially resembles a foramen ovale in which the valve has failed to form, if the specimen is held up to the light, the de

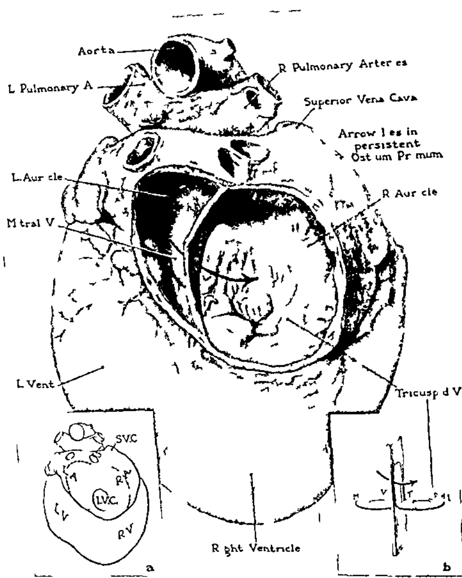


Figure xxiii 4 Persistent ostium primum Child
The m tral and tricusp d valves form the base of the defect

fect can usually be seen to bisect the true foramen ovale, which is outlined by a thin walled membrane imbedded in the auricular wall (see Figures xxiii–5 and 8) One of the essential features of this type of defect is that it lies in the auricular septal wall and is completely surrounded by a rim of tissue An auricular septal defect is often erroneously spoken of as a "patent ostium secundum" or a 'patent foramen ovale" The term *patent foramen ovale* should be restricted to those cases in which the foramen ovale is normally formed and is covered by a valve which is not completely sealed

The foramen ovale is normally patent at birth Studies by Barclay et al[1] indicate that functional closure of the foramen ovale occurs before the functional closure of the ductus arteriosus Functional closure of both pathways normally occurs shortly after birth Anatomical closure of the foramen ovale, however, does not occur for several months it is rare before two months of age and frequently is not complete until the second year of life Indeed, probe patency of the foramen ovale persists throughout life in 57 to 25 per cent of all persons [3] Hence such a condition should not be regarded as a malformation, but rather as a variant of the normal Inasmuch as the valve which closes the foramen ovale lies on the left side of the auricular septum, and consequently opens from right to left, as long as the pressure in the left auricle is equal to or slightly greater than that in the right auricle, the valve is functionally closed Such patency of the foramen ovale is of no functional importance as long as the normal auricular pressure relations are maintained Nevertheless, any malformation on the right side of the heart which raises the pressure in the right auricle above that of the left auricle will force the valve covering the foramen ovale to open Under such circumstances the foramen ovale acts as an escape valve for the high pressure in the right auricle but at the same time it permits the establishment of a right-to-left shunt

The condition becomes of clinical importance in malformations of the heart which place a strain on the right auricle Thus in isolated valvular pulmonary stenosis, primary pulmonary hypertension, and Ebstein's anomaly of the tricuspid valve, the increased pressure in the right auricle tends to hold the foramen ovale open Consequently patency of the foramen ovale occurs in approximately 75 per cent of patients with these malformations In such instances it is the right-to-left shunt through the foramen ovale which causes the cyanosis Cyanosis of this origin usually develops insidiously between two and seven years of age (see Chapters xvii–xix)

Patency of the foramen ovale may also become of clinical significance in con

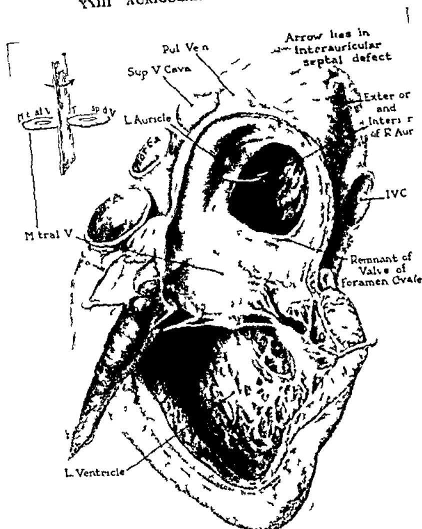


Figure xxiii–5 Auricular septal defect with mitral stenosis a pulmonary artery of normal size, and a small aorta Child

ditions which cause great enlargement of both auricles Under such circumstances the auricular wall becomes stretched, whereas the free margin of the valve which covers the foramen ovale does not become proportionally stretched Consequently a defect in the auricular septum is created This occurs most commonly in an individual who has had a severe rheumatic infection Under such circumstances the injury to the mitral valve usually causes the pressure in the left auricle to be abnormally high this sets in motion a sequence of events similar to that produced when there is a gross defect in the auricular septum (see Section B)

Occasionally patency of the foramen ovale is of clinical importance in that permits the occurrence of paradoxical emboli

The several types of defects in the auricular septum are discussed in the fo lowing sections Section A concerns a single auricle, Section B defects of the o tium secundum type, Section C defects of the ostium primum type, and Sectio D a persistent ostium atrioventriculare commune

A *A Single Auricle*

A single auricle means that the auricular septum has failed to develop an no wall separates the two auricles Such a defect is common when there is a isolated dextrocardia or a situs inversus with a levocardia In both instances th inferior vena cava lies on the same side of the body as the ventricles Conse quently the lines of stress and strain in the auricles are greatly altered, there i usually a gross defect in the auricular septum or the auricular septum is entirel absent

Although a single auricle may occur in tricuspid atresia, it is more commo to find a large defect in the auricular septum than a true single auricle

A single auricle is apparently the type of cardiac abnormality which is com mon in the Ellis–van Creveld syndrome,[4] namely, ectodermal dysplasia, poly dactyly, and chondrodysplasia A review of the literature shows that, in mos of the cases in which there was an associated cardiac abnormality, autopsy re vealed a single auricle The experience in our clinic confirms this observation Occasionally a single auricle exists in an otherwise normal heart

NATURE OF THE MALFORMATION

A single auricle means that the two atria constitute one large chamber into which the superior vena cava and the inferior vena cava open on the right side and the pulmonary veins enter in their normal position The mitral and tricuspid valves are normally formed and competent each opens into its respective ven tricle Both ventricles are normally formed and the ventricular septum is intact The pulmonary artery arises from the right ventricle and the aorta from the left ventricle The ductus arteriosus undergoes normal obliteration

COURSE OF THE CIRCULATION

Most of the blood from the right side of the common auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs, where it is oxygenated, and is returned in the normal fashion by the pulmonary

veins to the left side of the common auricle In the common auricle there is free admixture of venous and arterial blood Hence some oxygenated *blood is directed to* the right side of the heart and some venous blood *to the left* This admixture of arterial and venous blood flows into the left ventricle and is pumped out through the aorta to the systemic circulation and *returned by the superior* vena cava and the inferior vena cava to the right side of the common auricle There the cycle starts again (see Diagram XXIII-1)

PHYSIOLOGY OF THE MALFORMATION

Inasmuch as there is free admixture of venous and arterial blood in the common auricle and, moreover, the oxygen saturation of the venous blood is normally 70 per cent, the oxygen saturation of the mixed venous and arterial blood tends to be well over 80 per cent Furthermore, the right auricle is a more readily distensible chamber than is the left and the pressure in the right auricle is normally lower than that in the left, consequently, although there is free admixture of venous and arterial blood in the common auricle, and although the direction of the shunt can readily change, the right side of the heart bears the burden There is a greater shunt from left to right than from right to left The right side of the common auricle and the right ventricle become dilated and hypertrophied The increased pulmonary blood flow means that a large volume of oxygenated blood is mixed with a smaller volume of venous blood, there is no visible cyanosis Inasmuch as the shunt is at the auricular level, the right ventricle has to pump a larger volume of blood than normal and the right ventricular pressure may be slightly elevated The pulmonary vascular bed is normal and the lungs can usually readily accommodate the increased volume of blood Hence, although the pulmonary pressure may be slightly elevated, it is usually less than *the pressure in the right ventricle* Furthermore, inasmuch as there is but a single auricle, great variation in the volume of blood directed to the *respective ventricles can readily* occur Hence there is real danger of upsetting the balance of the *circulation*

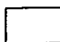
CLINICAL FINDINGS

The clinical findings are closely similar to those in any patient with a large *left-to-right shunt*

Left sided chest deformity is the rule, as the right *side of the heart* bears the brunt of the abnormal circulation and the right *side* of the common auricle and the right ventricle *become dilated* and *hypertrophied*

Diagram XXIII–1

Head and upper extremities
R Lung
P.A.
S.V.C.
PV
Aorta
Pul. A
L Lung
Pul. V
Pulmonary Artery
L.A.
R.A.
L.V.
R.V.
Descending Aorta
I.V.C.
Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis

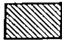

Venous blood

DIAGRAM XXIII-1

Single auricle

The fundamental feature of this malformation is that the auricular septum is absent and the two auricles function as a single chamber

The blood from the right side of the common auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs, where it is oxygenated, and returned in the normal fashion by the pulmonary veins to the left side of the common auricle. From there most of the blood flows into the left ventricle and is pumped out through the aorta to the body and returned through the superior vena cava and the inferior vena cava to the right side of the common auricle.

The musculature of the right auricle is more readily distensible than that on the left side. Therefore anything which raises the pressure on the left side establishes a predominant left to-right shunt. Consequently an increased volume of blood is pumped out to the lungs and returned to the left auricle. This in turn raises the pressure on the left side of the common auricle and again blood is shunted to the right side So the cycle continues Nevertheless, the absence of an auricular septum means that some blood is shunted *from the right side to the left There is usually slight oxygen un*saturation of the arterial blood

Clinical diagnosis The patient may have a frail build He may show slight cyanosis, which varies from day to day The heart may be slightly enlarged and tends to undergo progressive enlargement A precordial systolic murmur is common and there may be a low pitched mid diastolic murmur Episodes of acute cardiac failure are not uncommon The x ray shows fullness of the pulmonary conus and increased hilar shadows The electrocardiogram shows a right axis deviation and a slight tendency toward right ventricular dominance

Diagram XXIII-1

Arterial blood (fully saturated)



Small admixture of venous blood
No visible cyanosis

Venous and arterial blood
Cyanosis visible

Venous blood

from the common auricle may be directed into either ventricle Anything which raises the pressure on the left side of the heart will cause an excessive volume of blood to flow into the right side of the auricular chamber Under such circum stances the patient may develop right sided cardiac failure Fortunately the child usually regains compensation quite as readily as he develops decompensation Indeed, he usually regains compensation with simple rest in bed, without the use of digitalis or of a diuretic

X RAY AND FLUOROSCOPIC FINDINGS

The heart is slightly enlarged to the right and to the left The pulmonary conus is full and hilar markings are increased (see Figure xxiii-6) In the left anterior-oblique position the right ventricle is seen to be enlarged In the right anterior-oblique position there is no enlargement of the left auricle.

Upon fluoroscopy a hilar dance may be visible

ELECTROCARDIOGRAPHIC FINDINGS

Usually the standard leads show a slight right axis deviation and the unipolar precordial leads show evidence of right ventricular hypertrophy



FIGURE XXIII-6 Single auricle Child

Cyanosis is usually absent Transitory episodes of cyanosis are, however, relatively common Thus cyanosis may develop upon exercise or may occur without any precipitating cause Some days the infant or child may show slight cyanosis of the lips and of the fingers and toes, and other days his color is entirely normal Cyanosis usually becomes apparent during an episode of cardiac failure

Clubbing of the extremities is absent, as the oxygen unsaturation of the arterial blood is but slight

Pallor may be conspicuous but in spite of this the red blood cell count, the amount of available hemoglobin, and the hematocrit reading are usually at high normal levels This combination of findings should always suggest the possibility of slight unsaturation of the arterial blood

Growth and development are basically normal In the outstanding example of this malformation which the author has had opportunity to study, the child was considered to be the healthiest of four children

Exercise tolerance is, however, slightly limited These children seldom can keep up with their coevals at play Nevertheless, they do not squat when tired

Pulmonary congestion is relatively common because of the increased pulmonary blood flow For this reason there may be an increased susceptibility to bronchitis and pneumonia

The liver may or may not be enlarged and may vary in size from day to day If cardiac failure occurs precipitously the liver becomes rapidly engorged and is extremely tender Indeed, the sudden onset of abdominal pain may be the first evidence of cardiac failure

CARDIAC FINDINGS

The heart may be normal in size or slightly to moderately enlarged The contour of the heart is similar to that which occurs in a partial anomaly of the pulmonary venous return and in an auricular defect of the ostium secundum type

The pulmonic second sound is widely split and hence seems accentuated

A systolic murmur may be widely heard over the precordium and a *low pitched mid diastolic murmur,* such as is frequently heard with a poorly functioning heart, is common

• *Progressive cardiac enlargement* usually occurs slowly over a period of years because of the easy reversal in the volume of the shunt in either direction

Episodes of cardiac failure may occur suddenly without great cardiac enlargement Such episodes are undoubtedly due to the ease with which the blood

slight oxygen unsaturation of the arterial blood The contour of the heart is, however, different in that in the malformation under discussion there is evidence of enlargement of the "right" auricle, the right ventricle, and the main pulmonary artery Cardiac catheterization shows that the shunt is at the auricular, not the ventricular, level

An Eisenmenger complex may also be considered because of the evidence of a predominant shunt from left to right and slight cyanosis Cardiac catheterization readily differentiates the two, as in the Eisenmenger complex the shunt is at the ventricular level and the pulmonary pressure is elevated

COMMONLY ASSOCIATED MALFORMATIONS

Isolated dextrocardia, situs inversus with levocardia, and a single ventricle are the most commonly associated malformations

A functional single auricle may occur with tricuspid or mitral atresia

The condition has also been reported in patients with pulmonary stenosis with or without a tetralogy of Fallot

TREATMENT

The surgical correction of this malformation is more difficult than that of an auricular septum of the ostium secundum type Nevertheless, in the only such patient for whom the author has recommended operation, Dr Henry Bahnson was able to invaginate the auricular wall and approximate the upper and lower portions he thereby constructed an auricular septum without the insertion of a patch

PROGNOSIS

The prognosis without operation is relatively good, as cardiac enlargement occurs extremely slowly Operation can restore the circulation to normal and thus render the prognosis excellent

SUMMARY

A single auricle may occur as an isolated malformation When there is but a single auricle, there is free admixture of venous and arterial blood in the common auricle but the predominant shunt is from left to right A large volume of blood flows to the lungs and is returned to the common auricle Hence there is usually no visible cyanosis Nevertheless, there is always slight oxygen unsaturation of the arterial blood Furthermore, the ease with which the volume of the shunt may be altered may cause episodes of transient cyanosis

SPECIAL TESTS

The oxygen saturation of the arterial blood is always slightly reduced, as there is free admixture of venous and arterial blood in the common auricle

Cardiac catheterization reveals a marked increase in the oxygen content of the blood in the "right" auricle as compared with that in the superior vena cava Indeed, the oxygen content may be so high as to suggest an anomaly of the pulmonary venous return The pressure in the right ventricle may be slightly elevated but that in the pulmonary artery is usually lower than that in the right ventricle

Angiocardiography may show a remarkable separation in the two streams of blood Thus in the lateral view it may appear that there is an auricular septum Furthermore, the swirling of dye within the auricle may give the appearance of a shunt in either or in both directions A selective angiocardiogram with the dye injected into the left side of the auricle may show the absence of the auricular septum

DIAGNOSIS

The diagnosis is based upon the finding of right sided cardiac enlargement with fullness of the pulmonary conus and evidence of a left to-right shunt in a patient who has slight oxygen unsaturation of the arterial blood and who shows occasional slight cyanosis or suffers from episodes of decompensation

DIFFERENTIAL DIAGNOSIS

The condition requires differentiation from anomalies of the pulmonary venous return, from an auricular septal defect of the ostium secundum type, from a single ventricle, and occasionally from an Eisenmenger complex

A partial anomaly of the pulmonary venous return is a common error in diagnosis because of the right sided enlargement and evidence of a left to-right shunt and the marked increase in the oxygen saturation of the arterial blood in the right auricle The fact that the arterial blood is not fully saturated differentiates the malformation under discussion from a partial anomaly of the venous return

An auricular septal defect of the ostium secundum type shows the same differential feature The arterial blood is fully saturated in this type of auricular septal defect, whereas in the malformation under discussion there is slight unsaturation

A single ventricle may be considered because of the slight cyanosis and the

toms produced by an auricular septal defect will vary greatly throughout the world In the areas where rheumatic fever is severe and rheumatic heart disease is common [illegible] rare, patients with auricular septal defects [illegible] enlargement In the United States and in western Europe, [illegible] defects and where rheumatic fever is inter[illegible] decreasing [illegible] seek medical advice because their child has slight [illegible] children will come to the doctor when there is little or no appreciable cardiac enlargement Nevertheless, the frequent association of this malformation with rheumatic fever is important to remember Furthermore, the signs of rheumatic heart disease may occasionally overshadow those produced by the malformation

NATURE OF THE MALFORMATION

An auricular septal defect of the so-called ostium secundum type is usually relatively large, 2 to 3 cm in diameter Its location is subject to wide variations, it may be anywhere in the auricular septum Not infrequently it occupies a position close to the foramen ovale The defect resembles the foramen ovale in that the wall of the auricular septum surrounds the defect It differs from probe patency of the foramen ovale in that the opening is not covered by a valve or membrane Occasionally, if the valve covering the foramen ovale is not completely sealed, great enlargement of the auricles may stretch the wall to such an extent that a defect is created Whenever there is a gross defect in the auricular wall, there is free communication between the two auricles (see Figures XXIII–5 and 8)

The condition becomes of clinical importance whenever there is a significant difference in the pressure between the two auricles Normally the pressure in the left auricle is slightly greater than that in the right auricle and consequently a left to-right shunt is established This malformation may be associated with an abnormality of the mitral valve—either a congenital malformation or an acquired mitral stenosis Regardless of the etiology, stenosis of the mitral valve tends to raise the pressure in the left auricle and increases the left to-right shunt Under such circumstances this malformation places a great strain on the right side of the heart The right auricle, the right ventricle, and the pulmonary artery are enlarged The left ventricle and the aorta are relatively small Indeed, it is usual to find the pulmonary artery twice the size of the aorta If the pulmonary artery is of normal size, the aorta is abnormally small if the aorta is of normal size, the pulmonary artery is huge

Lutembacher s syndrome [6] is an auricular septal defect combined with a

The heart may or may not be enlarged The pulmonic second sound is accentuated A precordial systolic murmur is the rule The condition leads to slow, progressive cardiac enlargement Even in the absence of great cardiac enlargement, episodes of acute cardiac failure may occur

The x ray contour of the heart is similar to that in other cases of a left to right shunt The right side of the auricle and the right ventricle are enlarged There is fullness of the pulmonary conus and the vascularity of the lungs is increased

The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy Cardiac catheterization shows that there is a left to right shunt at the auricular level and that the arterial blood is not fully saturated

The diagnosis is suggested by findings of right sided cardiac enlargement with a precordial systolic murmur and an accentuation of the pulmonic second sound in a patient who shows episodes of transient cyanosis The patient may suffer from episodes of acute cardiac failure without great cardiac enlargement

The condition calls for differentiation from an auricular septal defect of the ostium secundum type, from anomalies of the pulmonary venous return, from an Eisenmenger complex, and also from a single ventricle with increased pulmonary blood flow

The surgical correction of this malformation is now possible and changes the prognosis from good to excellent

B *Auricular Septal Defects of the Ostium Secundum Type and Lutembacher's Syndrome*

Defects in the auricular septum of the ostium secundum type are relatively common When such a defect occurs as an isolated malformation, it is extraordinarily well tolerated in childhood Individuals with an auricular septal defect are, however, unusually susceptible to pneumonia and may be susceptible to rheumatic fever, furthermore, they are prone to develop all sorts of cardiac arrhythmias It is frequently because of such complications that patients seek medical attention An uncomplicated auricular septal defect produces almost as few physical findings as symptoms Nevertheless, over a period of years, the signs and symptoms produced by this malformation are greatly altered if the patient develops rheumatic heart disease or essential hypertension Indeed, it is a striking fact that, with the decrease in the frequency of rheumatic fever and with the great reduction in the severity of rheumatic heart disease, the signs and symptoms which occur in patients with an auricular septal defect have greatly changed For this reason it seems probable that for many years the clinical symp

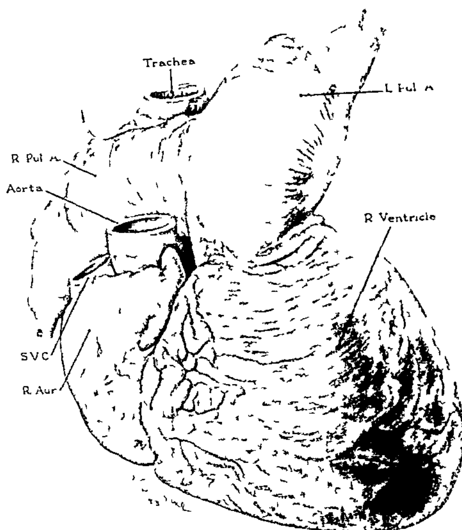


FIGURE XXIII–7 Lutembacher s complex showing enormous dilatation of the pulmonary artery (same patient as in Figure XXIII–8) Adult

congenital or acquired mitral stenosis and enormous dilatation of the pulmonary artery, as shown in Figures xxiii–7 and 8 The abnormal size of the pulmonary artery is an integral part of the malformation Mitral stenosis, either congenital or acquired, increases the strain on the right side of the heart and increases the dilatation of the pulmonary artery It is the great enlargement of the pulmonary artery which differentiates this condition both clinically and at autopsy from other auricular septal defects The enormous size of the pulmonary artery gives the heart a characteristic contour (see Figures xxiii–7, 12, and 13) Nevertheless, the circulation of the blood, the physical findings, and the clinical course are essentially the same regardless of the size of the pulmonary artery

Owing to the fact that as the auricles enlarge the auricular septum becomes stretched in all directions, defects in the auricular septum may increase in size as the patient grows Consequently it is not unusual in adults to find an auricular septal defect which is so large that the heart of a newborn baby could be passed through it

COURSE OF THE CIRCULATION

During fetal life circulation to the lungs is minimal, the ostium primum and the ostium secundum represent the normal fetal pathways, the flow of blood through the foramen ovale is from right to left It is not until after birth, with the establishment of the pulmonary circulation and with the increased volume of blood returned from the lungs to the left auricle, that the valve which covers the foramen ovale closes Therefore the defect in the auricular septum places no strain on the fetal circulation At birth the heart is normal in size

When this malformation is associated with a congenital mitral stenosis, the mitral stenosis alters the course of the fetal circulation Under such circumstances blood will flow less easily from the left auricle to the left ventricle and the pressure in the left auricle will be abnormally high Consequently less blood will flow from the right auricle to the left auricle and more blood will flow from the right auricle into the right ventricle and out by way of the pulmonary artery and thence through the ductus arteriosus to the descending aorta Hence the work of the right side of the heart is increased, at birth the right auricle and the right ventricle and the pulmonary artery are larger than normal

At birth and in early infancy the pressure in the right auricle may be greater than that in the left auricle, and if so there will be a right to-left shunt For this reason the infant may be a ' blue baby or show cyanosis for a period of several days Occasionally the cyanosis is so persistent that it is thought it will be perma

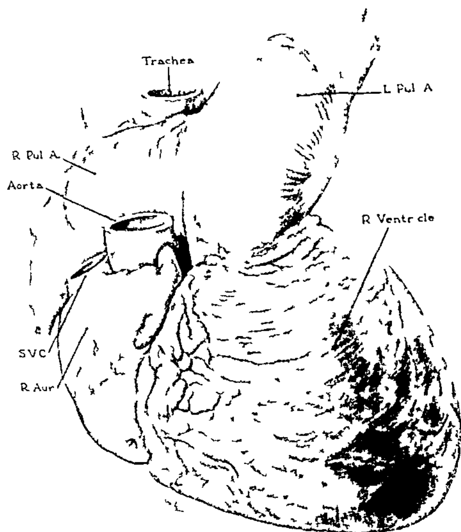


FIGURE XXIII–7 Lutembacher's complex showing enormous dilatation of the pulmonary artery (same patient as in Figure XXIII–8) Adult

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of the pulmonary artery to the lungs, and back by the *pulmonary veins* to the left auricle Thereupon the increased amount of blood which has been returned to the left auricle again raises the pressure in that chamber and blood is again shunted into the right auricle (see Diagram XXIII-2) The vicious cycle thus set up causes enlargement of the right auricle, the right ventricle, and the pulmonary artery, whereas the left ventricle and the aorta are spared

PHYSIOLOGY OF THE MALFORMATION

The normal pressure in the left auricle is slightly greater than that in the right auricle Therefore the shunt is from left to right Furthermore, any condition which raises the pressure in the left auricle, such as rheumatic heart disease or hypertension, increases the left to-right shunt As rheumatic fever becomes better controlled, hypertensive heart disease will become increasingly important as a factor which exaggerates the left to-right shunt and initiates difficulty

The left to-right shunt through the defect in the auricular septum causes the pulmonary circulation to receive more than its normal quota of blood and the systemic circulation to receive less Consequently the body is starved and the lungs are excessively vascular and *pulmonary congestion is common* The right ventricle is required to pump an excessively large volume of blood, it becomes dilated and slightly hypertrophied The pressure in the right ventricle is slightly to moderately elevated Nevertheless, severe pulmonary hypertension seldom develops Usually the pressure in the pulmonary artery is found to be slightly *lower than* that in the right ventricle Indeed, it is common to find that the pressure in the pulmonary artery is 10 to 20 mm of mercury lower than that in the right ventricle without organic pulmonary stenosis

SEX INCIDENCE

It is a curious fact that this malformation occurs more frequently among females than among males Although the explanation is not known, the *ratio of* incidence is two *to one*[7 8]

CLINICAL FINDINGS

At birth and in infancy the clinical findings are subject to wide variation It is quite as frequently the associated illnesses *as it is the cardiac* symptoms which bring the infant to the doctor The occurrence of some cardiac arrhythmia, especially paroxysmal *tachycardia*, during the *first months* of life, suggests the malformation under discussion Not infrequently *great difficulty* is encountered in

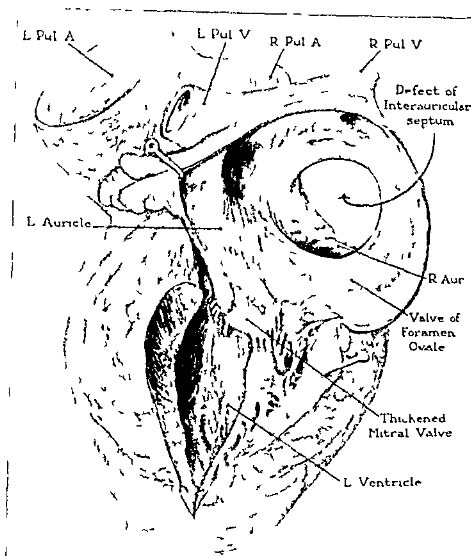


FIGURE XXIII–8 Lutembacher's complex showing an auricular septal defect, enormous dilatation of the pulmonary artery, and mitral stenosis (same patient as in Figure XXIII–7) Adult

nent It is only with the establishment of the normal pressure relations between the systemic and pulmonary circulations that a reversal in the direction of the shunt occurs and cyanosis disappears

In later life any condition which raises the pressure in the left auricle—such as a congenital mitral stenosis, a rheumatic infection with insufficiency or stenosis of the mitral valve or the aortic valve, or hypertension—places a strain on the left side of the heart and starts a vicious cycle The increased pressure in the left auricle forces the blood from the left auricle into the thin walled, readily distensible right auricle Thence the blood passes to the right ventricle, out by way

Diagram XXIII–2

Auricular septal defect of the ostium secundum type

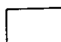
The fundamental feature of this malformation is a gross defect in the auricular septum, which may or may not occupy the position of the foramen ovale. The defect becomes of functional importance whenever the pressure in the two auricles is unequal In early infancy the pressure may be greater in the right auricle than in the left, if so, the shunt is from right to left. With the growth of the individual and the various vicissitudes of life, the pressure in the left auricle rises When the pressure in the left auricle exceeds that in the right, a chain of events is set in motion which leads to a characteristic clinical syndrome.

The blood from the right auricle passes into the right ventricle and out by way of the pulmonary artery to the lungs The oxygenated blood from the lungs is returned by the pulmonary veins to the left auricle Most of the blood from the left auricle flows in the normal fashion into the left ventricle and is pumped out by way of the aorta to the systemic circulation, this blood is returned by the superior and inferior venae cavae to the right auricle Because of the high pressure in the left auricle, part of the blood from the left auricle is shunted through the defect in the auricular septum to the right auricle The blood so shunted passes into the right ventricle, thence it is pumped out through the pulmonary artery to the lungs and is again returned by the pulmonary veins to the left auricle Thus an excessive amount of blood is pumped around and around the pulmonary circulation whereas the left ventricle, aorta, and systemic circulation receive less than their normal quota of blood The right auricle and right ventricle are enlarged The pulmonary artery is usually twice the size of the aorta The strain on the left auricle is relieved by the defect in the auricular septum Therefore, the left auricle is not enlarged The left ventricle is relatively small

Clinical diagnosis The patient has a frail build and poor physical development. Left sided chest deformity is the rule. There may be slight acrocyanosis but, inasmuch as the shunt is from left to right, there is no cyanosis and no clubbing

The murmurs are subject to wide variation The most characteristic findings are a precordial systolic murmur and a widely split second sound The systolic murmur is even better heard in the interscapular region than over the precordium The x ray shows that the heart is enlarged to the right and to the left and fullness of the pulmonary conus and fluoroscopy reveals a hilar dance but no evidence of left auricular enlargement The electrocardiogram shows a tendency to a right axis deviation and evidence of an incomplete right bundle branch block

DIAGRAM XXIII–2



Arterial blood (fully saturated)



Venous and arterial blood
Cyanosis visible

Small admixture of venous blood
No visible cyanosis



Venous blood

combination of these findings may be so striking that the diagnosis can be made at sight

A tracheal tug may be present, it is caused by the enormous size of the pulmonary artery and the resultant pressure on the trachea

The pulse pressure is narrow This is caused by the great left to-right shunt and the marked reduction in the volume of blood which reaches the systemic circulation For the same reason the *blood pressure* is low

Clubbing and *cyanosis* are usually absent, as the shunt is normally from left to right Not infrequently there is slight acrocyanosis This is usually due to the increased deoxygenation of the blood in the peripheral tissues associated with the diminished blood volume and the increased utilization of the available oxygen by the body Since the major shunt is from left to right, there is never the intense cyanosis and concomitant clubbing characteristic of a predominant venous-arterial shunt unless there is some additional abnormality such as primary pulmonary hypertension

Transitory or terminal reversal in the direction of the shunt is more common in this condition than in any other malformation, because the difference in the relative pressure in the two auricles is slight and the defect in the auricular septum is large A reversal in the direction of the shunt may cause intense cyanosis but is seldom of sufficient duration to produce clubbing If deep, persistent cyanosis and clubbing are present, it is indicative of some additional complicating factor

CARDIAC FINDINGS

The heart is usually enlarged and overactive *A precordial heave* is common, especially in older individuals, it is indicative of a right ventricle which is carrying an excessive load The *apical thrust* can be seen to be displaced downward and outward it often lies in the sixth interspace in the anterior axillary line The *cardiac dullness* is increased to the right as well as to the left, owing to the great enlargement of the right auricle

The pulmonic second sound is loud and widely split The reduplication of the second sound is caused by the asynchronous closure of the aortic and pulmonic valves The relative time of the closure of these two valves is unaltered by exercise Consequently the splitting of the second sound remains constant, for this reason it is frequently spoken of as fixed The increased blood flow through the pulmonary circulation frequently renders the action of the pulmonary valve so forceful that its closure is readily palpable

getting the infant to gain weight Sometimes the infant will scarcely gain any weight for the first six months of life and in the second six months will gain as a newborn baby should In addition, these infants are usually prone to respiratory and pulmonary infections Repeated attacks of pneumonia menace their lives Barring such accidents, in spite of murmurs and thrills, and in some instances considerable cardiac enlargement, these babies survive

The condition may be asymptomatic Suspicion may first be aroused when an x ray of the chest, taken for some entirely different reason, shows an abnormal contour of the heart

Frequently the true nature of the condition is not recognized until after the subsidence of a rheumatic infection, because the characteristic clinical syndrome does not develop until some condition places sufficient strain on the left auricle to establish a significant left to-right shunt

Thus, although this malformation is present from birth, the physical signs and symptoms make their appearance with the growth and development of the individual in combination with intercurrent illnesses and the stress and strains of life Consequently, although diagnosis may be difficult in infancy, with the passage of years the majority of patients with this malformation develop a distinctive clinical syndrome

The various illnesses, especially superimposed rheumatic infection, and the elevation of the blood pressure so common with advancing years, increase the strain on the left side of the heart, thus augment the left to-right shunt, and thereby render the clinical picture more characteristic

A superimposed rheumatic infection has a threefold effect upon the size of the heart First, the infection of the myocardium causes cardiac enlargement Second, the progressive stenosis of the mitral valve increases the shunt from left to right and actually places an additional load on the right side of the heart Third, with the enlargement of the auricles, the auricular septum becomes stretched, this stretching enlarges the size of the defect, which in turn augments the volume of the shunt and the load upon the heart Thus, as the patient grows older, the size of the defect tends to become greater Indeed, the malformation is one of the few compatible with relative longevity in which repeated episodes occur that lead to progressive cardiac enlargement

The appearance of the individual is often characteristic [9] The patient has the frail build of the *gracile habitus* owing to the large left to right shunt Furthermore, the tremendous load on the pulmonary circulation leads to great enlargement of the right ventricle and causes marked *left sided chest deformity* The

and thrills Furthermore, the occurrence of valvular lesions usually intensifies the basal systolic murmur and thrill associated with the defect in the auricular septum

Lutembacher s complex, that is, a gross defect in the auricular septum combined with congenital or acquired mitral stenosis and enormous dilatation of the pulmonary artery (see Figures XXIII–7 and 8) produces a clinical syndrome which, although seldom seen today, is so distinctive that the diagnosis can frequently be made from the history and the appearance of the patient The history is one of repeated attacks of pneumonia, flu, grippe, or polyarthritis The patient has a *gracile habitus* with marked left sided chest deformity, a pronounced precordial heave, and an apical thrust which is frequently visible outside the mid-clavicular line On palpation a pronounced systolic thrill is palpable over the pulmonary area and both a systolic and a diastolic thrill are palpable at the apex. On auscultation a harsh systolic murmur is audible over the pulmonary area, in addition there is a systolic murmur which is maximal at the apex and transmitted to the axilla and also a long, low pitched, mid-diastolic murmur which is best heard within the apex Indeed, when the murmur and thrill over the base of the heart suggestive of a congenital malformation are combined with signs at the apex characteristic of rheumatic heart disease, the possibility of Lutembacher s syndrome should be considered

An early diastolic murmur may sometimes be audible along the left sternal border It is due to relative pulmonary insufficiency, hence it is more frequently heard in older individuals than in young children

Murmurs may not be noted until the terminal illness The author has also found the reverse to be true murmurs which have been present may disappear As the pressure in the lesser circulation increases, the difference in pressure between the two circulations becomes less, this tends to diminish the intensity of the murmurs and thrills

Auricular fibrillation may also cause the murmur to disappear, as in the two cases reported by Cossio and Arana [11]

It is important to remember that, although in some cases the murmurs are extremely characteristic, too much emphasis should not be placed on auscultatory findings Indeed, the murmurs and thrills may be so insignificant as to render the diagnosis doubtful

Cardiac failure is usually a late manifestation Cardiac failure may be caused by a superimposed rheumatic infection, consequently as the infection subsides the patient is able to regain compensation In the United States and in other

Murmurs and *thrills* are subject to considerable variation They are pro by the malformation and may be complicated by a superimposed rheuma fection The murmurs depend upon the relative pressure in the greater an lesser circulation In early infancy there may be no murmur or there may loud systolic murmur and a low pitched mid diastolic murmur To the b the author's knowledge there is nothing in early infancy to differentiate murmurs from those caused by a patent ductus arteriosus or by a ventri septal defect Nevertheless, the findings of great cardiac enlargement, x ra dence of fullness of the pulmonary conus, and congestion in the lungs i absence of a murmur should always suggest the possibility of an auricular s defect Thus the absence of a murmur, but not its presence, may aid in the d entiation of this condition from persistent patency of the ductus arterios from a ventricular septal defect

As the patient grows, the pressure in the left auricle tends to rise Whe pressure in the left auricle becomes significantly greater than that in the auricle, a systolic murmur is usually audible to the left of the sternum ir second and third left interspaces This murmur is often not very loud and readily be confused with a functional murmur Although on a theoretical there is every reason to believe that a murmur which originates from an au lar defect is presystolic in time, insofar as the human ear can differentiate, murmur is systolic in time Many theories[10] have been advanced to explain occurrence of the systolic murmur It is the author's impression that this mur probably originates in auricular diastole which coincides with early ventric systole It is possible that the murmur has its origin in the right ventricle o the pulmonary artery, but its intensity cannot be correlated solely with the of the pulmonary artery

The systolic murmur is often well heard posteriorly, high in the interscap region In many instances the murmur is louder posteriorly than anteriorly c the precordium If the murmur has a harsh, rasping quality and is well he along the left sternal border, the possibility of a persistent ostium primum w a cleft in the mitral valve or a large high ventricular septal defect should alw be borne in mind (see below, Section c, and Chapter XXIV, Section B)

A thrill over the pulmonary area is palpable when the murmur is inte This is far more frequently present in Lutembacher's syndrome than in an complicated auricular septal defect

Superimposed rheumatic infections may injure either the mitral valve or aortic valve or both These valvular lesions produce their characteristic murm



FIGURE XXIII-9 Auricular septal defect Infant

ous, it is not as pronounced as that seen in an anomaly of the pulmonary venous return

Examination in the left anterior-oblique position confirms the impression that the enlargement is mainly right sided. In this position the right ventricle is usually seen to project to the anterior chest wall The enlargement of the right ventricle is often so great as to displace the left ventricle backward, thus it appears to share in the enlargement

In the right anterior-oblique position the great enlargement of the heart causes the entire esophagus to be pushed backward, as shown in Figures XXIII-12 and 13 It is, however, unusual to see specific enlargement of the left auricle even in the presence of a mitral stenosis, because, as previously mentioned, the right auricle, not the left, bears the brunt of the strain caused by the mitral stenosis

ELECTROCARDIOGRAPHIC FINDINGS

In an auricular septal defect of the ostium secundum type the standard leads characteristically show a slight right axis deviation and the unipolar precordial leads show the pattern of an incomplete right bundle branch block or an RsR' pattern without prolongation of the QRS complex (see Figure XXIII-14) Evidence of marked right ventricular hypertrophy is usually not present because the enlargement of the right ventricle is not due primarily to hypertrophy but to a combination of dilatation and hypertrophy caused by the excessive volume of blood which is pumped around the lesser circulation

countries where the incidence of rheumatic fever has decreased and surgical correction of malformations of the heart is common, the occurrence of great cardiac enlargement or evidence of cardiac failure is seldom seen Nevertheless, some of the largest hearts compatible with life occur in persons with auricular septal defects and partial anomalies of the pulmonary venous return

When cardiac failure does occur, there is congestion in the lungs, engorgement of the liver, and edema of the extremities In addition, there may be changes in the quality and the intensity of the murmur and the thrill Frequently the findings in the heart resemble those of a poorly functioning rheumatic heart

X RAY AND FLUOROSCOPIC FINDINGS

The x ray is often of diagnostic aid [8, 10, 14] The distinctive features are the enlargement of the right auricle and the right ventricle, the prominent pulmonary conus, the small aortic knob, and the increased hilar shadows It is important to appreciate that there are two distinctive x ray contours, depending upon whether or not the pulmonary artery is greatly dilated

The usual finding is that of slight to moderate cardiac enlargement caused mainly by the dilatation and hypertrophy of the right auricle and the right ventricle The pulmonary artery is usually enlarged and the vascular markings are increased The prominence of the pulmonary conus may be conspicuous in infancy while the diaphragm is high (see Figure XXIII–9) and decrease as the diaphragm descends (see Figure XXIII–10) The prominence of the pulmonary conus usually again becomes conspicuous in adult life (see Figure XXIII–11)

Lutembacher's syndrome is differentiated from other defects of the ostium secundum type by the enormous dilatation of the pulmonary artery Not only is the main pulmonary artery huge, but one or both of the main branches of the pulmonary artery are also tremendously enlarged (see Figures XXIII–12 and 13) The enlargement of the pulmonary artery ends abruptly as the artery subdivides into its smaller branches The huge, dense shadow cast by the pulmonary artery and its branches may be so pronounced as to be mistaken for a mediastinal tumor In some cases the dilatation of the pulmonary artery appears to involve only the main stem Under such circumstances the pulmonary conus is enormous but there is no great increase in the vascular shadow to the right of the sternum

Upon fluoroscopy the tremendous mediastinal shadows are seen to pulsate, thus proving that shadows are vascular in origin Indeed, this malformation is one which causes a true dancing hilus, although the dance is usually conspicu

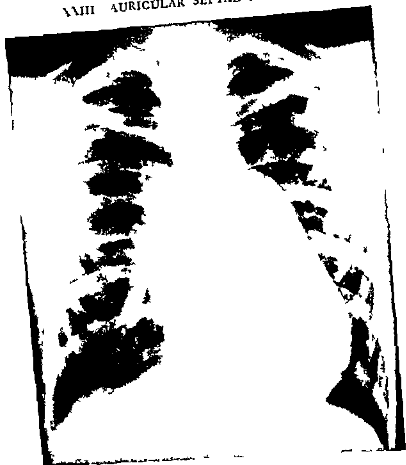


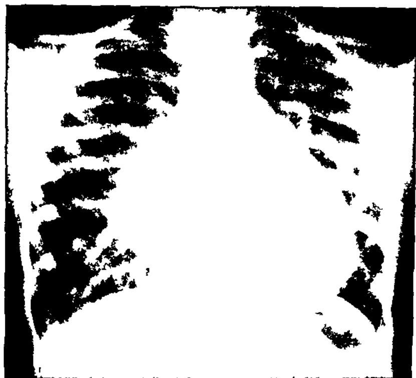
FIGURE XXIII-11 Auricular septal defect Adult

In Lutembacher s syndrome the standard leads frequently show prolongation of the P R interval and notching and widening of the QRS complex

The finding of a left axis deviation in the standard leads and evidence of left ventricular hypertrophy or even combined hypertrophy in the unipolar pre cordial leads is always suggestive of a defect of the ostium primum type (see be low)

SPECIAL TESTS

The red blood cell count, the amount of available hemoglobin, and the hema tocrit reading are normal or may be slightly below normal The erythrocytic sedimentation rate, in the absence of a superimposed infection, is normal The oxygen saturation of the arterial blood is normal

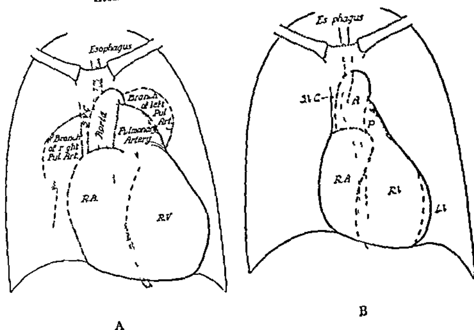


Before operation

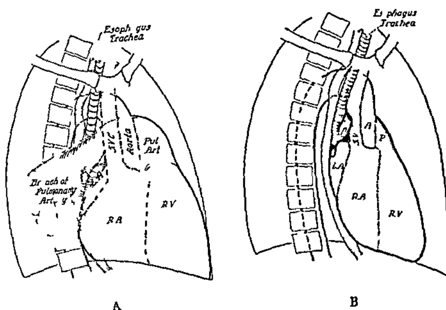


After operation

FIGURE XXIII-10 Auricular septal defect Child



ANTERIOR POSTERIOR POSITION



RIGHT ANTERIOR-OBLIQUE POSITION

FIGURE XXIII 13 (A) Lutembacher s syndrome with a dilated pulmonary artery and (B) normal heart Adult

Anterior
posterior
position

Right anterior
oblique
position

FIGURE XVIII-12 Lutembacher s syndrome showing enormous dilatation of the pulmonary artery Adult

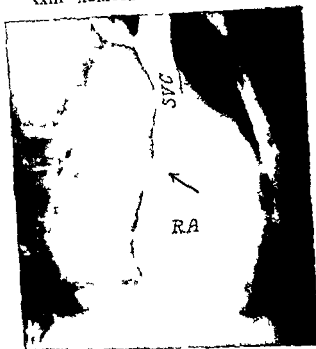


FIGURE XXIII–15 Auricular septal defect Adult
Arrow points to dye passing to the left auricle

ure XXIII–16 A defect which lies high up in the auricular septum close to the entrance of the superior vena cava may be clearly demonstrated in the lateral view Under such circumstances if the dye flows from the superior vena cava into both auricles, the auricular septum is thereby visualized and the existence of a defect high in the septal wall is proven (see Figure XXII–14) A defect in this locality is relatively common in association with a partial anomaly of the pulmonary venous return, but seldom, if ever, occurs in an auricular septal defect of the ostium secundum type Angiocardiography may also be of aid in that, after the return of blood to the left auricle, the late films may show such pronounced recirculation of the dye through the right side of the heart that the left ventricle and the aorta are poorly delineated

Selective angiocardiography may be of aid in the demonstration of an auricular defect if the catheter is passed into the left auricle and the dye is injected into that chamber In doubtful cases this procedure may aid in the differentiation between an ostium secundum defect and one of the ostium primum type In the former the lower rim of the auricular septum should be visualized by this technique

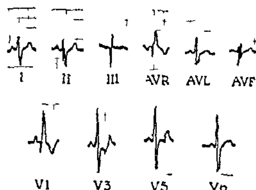


FIGURE XXIII-14 Auricular septal defect of the ostium secundum type

Cardiac catheterization shows an increase in the oxygen content of the blood in the right auricle as compared with that in the superior vena cava and that in the inferior vena cava The extent of the increase depends upon the size of the defect and the relation of the catheter to the stream of blood shunted from the left auricle to the right auricle It may or may not be possible to pass the catheter into the left auricle *The blood in the left auricle is fully saturated and so is* that in the left ventricle and the systemic circulation

There may or may not be a further increase in the oxygen content of the blood in the right ventricle as compared with that in the right auricle The pressure in the right ventricle is usually slightly elevated, the elevation of the pressure occurs when the volume of the shunt is enormous and the amount of extra work demanded of the right ventricle is greatly increased Nevertheless, it is a striking fact that the volume of pulmonary blood flow may be several times as great as that of the systemic blood flow and pressure in the right ventricle may be but slightly elevated The pressure in the pulmonary artery is generally directly proportional to or slightly less than that in the right ventricle

Unless the left auricle is catheterized there is nothing which clearly differentiates this malformation from an anomaly of the pulmonary venous return If the oxygen saturation of the blood in the right auricle rises abruptly to 97 or 100 per cent, it is always suggestive of an anomaly of the pulmonary venous return (see Chapter XXII, Section A)

Angiocardiography may also be of diagnostic aid In some cases it may be possible to see a jet of blood passing from the right auricle to the left auricle, as in Figure XXIII-15 *In other cases there is a slight swirl* of the dye in the right auricle as the blood which is shunted from the left auricle to the right auricle meets the dye injected through the superior vena cava into that chamber, as shown in Fig

tricle, fullness of the pulmonary conus, a small aorta, and increased hilar shadows which upon fluoroscopy are seen to pulsate

The electrocardiogram characteristically shows a slight right axis deviation and evidence of an incomplete right bundle branch block When the condition is complicated by superimposed rheumatic heart disease it is common to find widening and notching of the P waves

Cardiac catheterization offers confirmatory evidence by the demonstration of a shunt at the auricular level Angiocardiography may also show evidence of a shunt from left to right or from right to left

DIFFERENTIAL DIAGNOSIS

There are a number of conditions from which this malformation must be differentiated The commonest of these are anomalies of the pulmonary venous return, defects in other portions of the auricular septum, and high ventricular septal defects with increased pulmonary blood flow In infancy persistent patency of the ductus arteriosus may be confused with this malformation prior to the development of a continuous murmur In childhood, in addition to the above mentioned conditions, the malformation under discussion must be differentiated from idiopathic dilatation of the pulmonary artery, primary pulmonary hypertension, or even a Taussig Bing malformation combined with a partial anomaly of the pulmonary venous return In older patients the condition may require differentiation from rheumatic heart disease, pulmonary tuberculosis, mediastinal tumors, or substernal thyroid

Anomalies of the pulmonary venous return more closely simulate a gross defect in the auricular septum than does any other malformation

Difficulty in diagnosis arises when some or all of the pulmonary veins drain into the right auricle and there is a gross defect in the auricular septum Anomalies of the pulmonary venous return cause even greater difficulty than does an isolated defect in the auricular septum Therefore, if the heart continues to enlarge in the absence of any evidence of rheumatic infection, the possibility of anomalous drainage of some of the pulmonary veins into the right auricle should always be considered It is the vascularity of the lungs which above all else offers the clue to the correct diagnosis In an auricular septal defect the hilar markings may be so dense as to resemble a mediastinal tumor, nevertheless, they are more sharply demarcated and less diffuse than are those associated with an anomaly of the pulmonary venous return In the latter they are too diffuse for such a mistake to be made Angiocardiography may show a defect at the margin of the

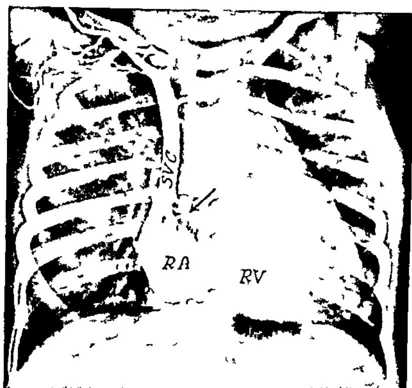


FIGURE XXIII–16 Auricular septal defect Child
Arrow points to dye filling defect

DIAGNOSIS

Diagnosis may occasionally be postulated from the history For example, in a woman known to have a congenital cardiac defect, who has had a large family and who has had repeated attacks of pneumonia, the most probable abnormality would be a gross defect in the auricular septum

Diagnosis is occasionally made at sight A frail, poorly developed individual, with conspicuous left sided chest deformity but with no cyanosis or clubbing, suggests the possibility of an auricular septal defect On physical examination the closure of the pulmonary valve is readily palpable in the second left interspace and only an insignificant systolic murmur may be audible When the second sound in this region is widely split and "fixed" and the murmur is better heard in the interscapular region than over the precordium, the malformation is probably an auricular septal defect The occurrence of a harsh systolic murmur and a thrill over the base of the heart, combined with signs of rheumatic heart disease at the apex, also is suggestive of this malformation

The contour of the heart in the x ray may suggest the diagnosis The characteristic findings are enlargement of the right auricle as well as of the right ven

sure is narrow A patent ductus arteriosus of such size as to give great cardiac enlargement and conspicuous pulsations in the lung fields is usually associated with a wide pulse pressure, a low diastolic pressure, and peripheral signs of aortic insufficiency In an auricular septal defect the electrocardiogram *usually shows* a right axis deviation and a right bundle branch block, whereas in a patent duc*tus arteriosus* a balanced axis and evidence of left ventricular hypertrophy are *the rule* An aortogram *is* the best means for the differentiation of the two conditions in infancy (see Chapter XX)

After the development of a continuous murmur the differentiation of these two conditions is easy In an auricular septal defect the basal murmur is limited to systole, whereas *in* a patent ductus arteriosus the murmur over the pulmonary area is continuous through systole and diastole

Idiopathic dilatation of the pulmonary artery may occur as an isolated anomaly Oppenheimer[15] has reported such a case The contour of the heart in the x ray closely resembles that of Lutembacher s syndrome When there are no murmurs or thrills, clinical differentiation of the two conditions may be impossible Usually, however, either the history or the examination of the heart will aid in the establishment of the correct diagnosis Special laboratory studies may be necessary Cardiac catheterization will show no change in the oxygen content of the blood in the right side of the heart and a normal pressure in the pulmonary artery

Primary pulmonary hypertension may require differentiation from an auricular septal defect, especially in those instances in which murmurs are not pronounced It is the marked accentuation of the pulmonic second sound which gives the clue to the diagnosis A faint systolic murmur and a loud early diastolic murmur are more common in patients with primary pulmonary hypertension than in those with an auricular septal defect The electrocardiogram usually shows a more pronounced right axis deviation and greater evidence of right ventricular hypertrophy in primary pulmonary hypertension than in an auricular septal defect Cardiac catheterization readily differentiates the two conditions (see Chapter XVIII)

A Taussig Bing malformation combined with a partial anomaly of the pulmonary venous return may show minimal cyanosis and right sided cardiac enlargement The cyanosis is, however, greater and the electrocardiogram usually shows evidence of more pronounced right ventricular hypertrophy in a Taussig Bing malformation than is ordinarily seen in an auricular septal defect Angiocardiography shows early dense opacification of the aorta immediately after the filling of the right ventricle

right auricle at the point of entrance of the pulmonary veins Cardiac catheteriza tion may show the blood at the margin of the right auricle to be fully or almost fully saturated

A single auricle may readily be confused with a gross defect in the auricular septum The occurrence of slight cyanosis of varying intensity and slight oxygen unsaturation of the arterial blood differentiates it from an auricular defect of the ostium secundum type

A defect of the ostium primum type, although a variant of an auricular septal defect, must be differentiated from a defect of the ostium secundum type The distinction is important because of the difference in surgical technique in the correction of the two malformations When there is a cleft in the mitral valve, the two conditions can usually be differentiated clinically, as the electrocardio-gram shows a tendency to a left axis deviation and evidence of combined hyper trophy (see Section c)

A persistent ostium atrioventriculare commune more closely resembles a ven tricular septal defect than an auricular septal defect, as the systolic murmur is usually harsh and rasping

A high ventricular septal defect with increased pulmonary blood flow may be mistaken for an auricular septal defect in infancy and childhood It is the finding of a harsh, rasping systolic murmur and a thrill along the margin of the left sternal border in the third and fourth interspaces, combined with a snapping second sound over the pulmonary area, which gives the clue to the diagnosis The electrocardiogram may show a right axis deviation but frequently shows a balanced axis or evidence of left ventricular hypertrophy A large ventricular septal defect is quite as incapacitating as is an auricular septal defect and in in fancy and childhood it does not produce cyanosis By late childhood or adoles-cence patients with an Eisenmenger complex usually show persistent cyanosis therefore this malformation causes greater difficulty in differential diagnosis in childhood than in adult life (see Chapter XXIV, Section B)

Persistent patency of the ductus arteriosus, prior to the development of a con tinuous murmur, may require differentiation from an auricular septal defect In infants and young children both malformations may be associated with great cardiac enlargement, a prominent pulmonary conus, and a dancing hilus An auricular septal defect may be associated with great cardiac enlargement and no murmur, whereas a ductus arteriosus sufficiently large to cause difficulty in early infancy almost invariably is associated with a harsh systolic murmur and a gal lop rhythm, even if there is no continuous murmur The pulse pressure is often of aid in the differential diagnosis In an auricular septal defect the pulse pres

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Rheumatic heart disease is a common complication and occasionally the signs of rheumatic heart disease may completely mask those of an auricular septal defect Simple awareness of the possibility aids in diagnosis Careful physical examination often reveals a systolic thrill over the pulmonary area Such a thrill should immediately arouse suspicion that the condition is not one of uncomplicated rheumatic heart disease When the defect lies in the lower part of the auricular septum and the basal systolic murmur is absent or so slight as to simulate a functional murmur, the existence of an underlying malformation may be difficult, if not impossible, to detect The picture is yet more complicated if auricular fibrillation is present Under such circumstances only a tentative diagnosis is possible on the basis of the characteristic cardiac contour, the tremendous right sided enlargement, and the absence of left auricular enlargement in the presence of mitral stenosis Cardiac catheterization may be necessary for a definitive diagnosis

Pulmonary tuberculosis is occasionally confused with an auricular septal defect In the author's experience this mistake is most common when the enormous dilatation of the pulmonary artery is limited to the main pulmonary artery and does not include the two primary branches Under such circumstances the congestion in the smaller branches of the pulmonary artery causes patchy hilar shadows These shadows are confused with parenchymal involvement of the lungs

The absence of any clinical or laboratory evidence of tuberculosis, combined with the cardiac contour and the occurrence of cardiac murmurs, differentiates these two conditions The demonstration of pulsations in the areas of suspected consolidation clinches the diagnosis of an auricular septal defect

Mediastinal tumors and Hodgkin s disease may be erroneously diagnosed when the hilar shadows are large and circumscribed As in tuberculosis, the fluoroscopic examination should clarify the nature of the pulmonary condition by the absence of pulsations in the hilar shadows when these are caused by tumors

Substernal thyroid[16] has been confused with the malformation under discussion because of pressure on the recurrent laryngeal nerve In an auricular septal defect x ray examination reveals the absence of a mass in the superior mediastinum and the presence of great cardiac enlargement

COMPLICATIONS

A superimposed rheumatic infection was formerly the most common complication In the earlier reports,[6, 8] 60 to 75 per cent of all persons with auricular

septal defects developed acute rheumatic fever The rheumatic manifestations may be so conspicuous as to mask the underlying congenital malformation and the condition is not infrequently mistaken for uncomplicated rheumatic heart disease The reverse is also true The signs of a congenital malformation may be so conspicuous as to overshadow completely the early signs of rheumatic myocarditis Frequently the patient who has obvious signs of a cardiac abnormality gives a history of acute rheumatic fever Because of the frequent association of these two conditions, in cases where the differential diagnosis lies between a congenital malformation of the heart and acquired heart disease, the possibility should be borne in mind of a rheumatic infection superimposed upon this particular malformation

Pneumonia and *pulmonary infections* are frequently encountered Patients with auricular septal defects are unusually susceptible to pneumonia Some have been known to have three and four attacks in a single year Indeed, it is not an uncommon experience for the doctor to be called because of pneumonia and upon physical examination to find evidence of great cardiac enlargement and a basal systolic murmur and thrill indicative of a congenital malformation In addition to pneumonia, these patients may suffer from repeated attacks of bronchitis and may even develop bronchiectasis

Cardiac arrhythmias of all forms are common Not only are extrasystoles frequent, but so are attacks of paroxysmal tachycardia, auricular flutter, auricular fibrillation, ventricular escape, and even complete dissociation These arrhythmias may develop during a rheumatic infection or a pulmonary infection, or without any obvious precipitating cause Inasmuch as cardiac arrhythmias frequently occur in association with great enlargement of one or both auricles, it is not surprising that they are common in auricular septal defects as the malformation causes great enlargement of the right auricle and is compatible with relative longevity

Pressure on the recurrent laryngeal nerve may result from the tremendous size of the heart and the great dilatation of the pulmonary artery Occasionally such pressure may cause a brassy cough In rare instances there may be complete paralysis of the left vocal cord or hoarseness similar to that produced by aneurysms of the arch of the aorta

Venous stasis and the formation of thrombi may occur as a late complication secondary to the great dilatation of the right side of the heart Thrombi may form in the right auricle, in the interstices of the right ventricle, and even in the pulmonary artery Portions of the thrombi may be thrown off into the lesser circulation, causing pulmonary infarcts Inasmuch as there is a gross defect in

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Pregnancy is remarkably well tolerated by women with this malformation, a number of them have had seven children[8] One is known to have had eleven Nevertheless, pregnancy greatly increases the work of the heart Large families are contraindicated

TREATMENT

Medical treatment is indicated if the patient suffers from cardiac arrhythmias, cardiac failure, or intercurrent infections

Cardiac failure should be treated with digitalis and the usual supportive measures Digitalis and quinidine are of value in the treatment of arrhythmias

The possibility of superimposed rheumatic infection should always be borne in mind If there is any evidence of an active rheumatic infection, this should be treated according to the best available medical knowledge Frequently after the subsidence of a rheumatic infection the patient will regain *compensation* and be able to resume a normal life

Continuous prophylaxis may be of great value The prophylactic administration of sulfonamides or of antimicrobial agents is often extremely beneficial in the prevention of bronchitis and pneumonia in infancy and early childhood prior to the correction of the malformation It is also indicated if the patient has had a rheumatic infection

Surgical closure of an auricular septal defect restores the heart and circulation to normal A number of methods for the closure of auricular septal defects have been developed The risk of operation is less than 5 per cent, therefore many persons desire surgery

Since the initial attempts at closure of an auricular septal defect by atrioseptopexy[19] or by use of a well,[9] many improvements have been introduced Søndergaard[1] showed that it was possible to dissect along the margin of the auricular septum in such a manner as to separate the two auricles sufficiently to place a purse string suture around the defect By this technique the defect can be closed without opening the auricles

Most surgeons prefer to operate under direct vision Bigelow s development of hypothermia opened up the possibility of temporary occlusion of circulation

the auricular septum, paradoxical emboli from the right auricle may occasionally cause cerebral accidents The author has seen two instances in which the enormously dilated pulmonary artery was virtually occluded by a thrombus This complication invariably causes obstruction in the pulmonary blood flow and consequently greatly increases the pressure in the right ventricle and it may lead to a reversal in the direction of the shunt Under such circumstances the clinical findings are completely altered, the patient develops cyanosis, which may lead to clubbing of the extremities and polycythemia

With the improvement in both medical and surgical treatment, all the above complications are becoming rare

Pulmonary hypertension occasionally develops in a patient with an auricular septal defect It is a serious complication but fortunately it is the exception not the rule Whether the pulmonary hypertension is secondary to the increased pulmonary blood flow, with or without the formation of emboli, or whether it is due primarily to some anomaly in the pulmonary vascular bed is not known Regardless of etiology, the development of pulmonary hypertension places an additional strain on the right ventricle, which is eventually transmitted back into the right auricle When the pressure in the right auricle rises sufficiently, a reversal in the direction of the shunt occurs This may be so great as to lead to the production of cyanosis, which tends to increase with exercise The development of cyanosis and polycythemia in a patient with an auricular septal defect is of extremely serious prognostic import

When pulmonary hypertension is extreme, it is probably primary, not secondary, to excessive pulmonary blood flow The author cared for one man with a gross defect in the auricular septum who in middle life developed both systemic and pulmonary hypertension The pulmonary hypertension was so severe that he suffered from a shunt reversal and developed cyanosis and polycythemia in spite of a systemic hypertension of over 200 mm of mercury Autopsy showed evidence of severe pulmonary vascular disease, but its etiology could not be determined

The late development of cardiac difficulties occasionally occurs after a relatively minor illness For example, a patient who has never been known to have any cardiac abnormality, after some slight illness may suddenly develop symptoms which lead to progressive cardiac failure Had such a patient been examined at an earlier date, it is probable that some evidence of a cardiac abnormality would have been detected

Subacute bacterial endocarditis, although notoriously common in patients with congenital malformations of the heart or with valves previously damaged

by a rheumatic infection, is extremely rarely associated with the malformation under discussion It is of great interest that in the only case the author has seen in which there was a proven Streptococcus viridans bacteremia or septicemia, autopsy showed no evidence of a subacute bacterial endocarditis[11] Nevertheless, subacute bacterial endocarditis has been known to occur in this malformation[1 18]

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Most surgeons prefer to operate under direct vision Bigelow s[2] development of hypothermia opened up the possibility of temporary occlusion of circulation

and thereby made operation possible on the interior of the heart without the necessity of a pump and oxygenator

Swan et al [23, 24] have perfected this technique and have used it in combination with coronary perfusion for the closure of auricular septal defects of the ostium secundum type In most instances the walls of the defect can be approximated and sutured together without the use of a prosthesis

Some surgeons prefer the use of a pump and oxygenator to hypothermia Each method has its advantage In the hands of experienced cardiovascular surgeons both methods give excellent results

The indications for surgery vary with the risk of operation and the amount of benefit to be gained from operation Some persons consider that the risk is so low that, once the diagnosis is established, the condition should be corrected It should, however, be remembered that small auricular septal defects are compatible with a long and active life, and that many women with this malformation have borne children with no undue difficulty Therefore the author does not feel that the mere presence of a small auricular septal defect is an indication for operation If there is evidence of increase in the size of the heart out of proportion to growth, operation is clearly indicated Evidence of cardiac enlargement or the occurrence of shortness of breath on exertion are also clear indications for operation Operation restores the heart and the circulation to normal Moreover, if there is cardiac enlargement, operation leads to a marked decrease in the size of the heart (see Figure XXIII–10) Therefore, if it seems probable that the patient may develop difficulty as a young adult, it is wise to operate before the patient assumes the responsibility of a family This is true for men as well as for women

The contraindications for operation are mainly associated conditions No patient should be operated upon during or immediately after an active rheumatic infection

Pulmonary hypertension adds greatly to the risk of operation For patients with such severe pulmonary hypertension that there is reversal in the direction of the shunt, closure of the defect is contraindicated

The complications from operation are remarkably few The two major complications are the occurrence of thrombi in the pulmonary vascular bed and the post pericardotomy syndrome

Older persons with great dilatation of the pulmonary arteries may suffer from thrombi either before operation or immediately after operation For this reason some physicians recommend the use of anticoagulants during the post operative period

Post pericardiotomy syndrome occurs more commonly following closure of an auricular septal defect than it does following other operations on congenitally malformed hearts The etiology is not clear but, if the patient shows evidence of a persistent pericarditis or a pleural rub, he should be kept quietly in bed until the inflammation has entirely subsided This simple precaution may obviate future difficulties

A persistent ostium primum will occasionally be confused with a defect of the ostium secundum type If the diagnosis of an auricular septal defect is based on the finding at cardiac catheterization of an increase in the oxygen content of the blood in the right auricle, an increasing number of patients will be found at operation to have a defect of the ostium primum type with a normal mitral and a normal tricuspid valve Unfortunately, when the base of the defect is formed by these valves, operation is difficult and usually requires the use of an extracorporeal circulation If such a condition is found at operation, the chest should be closed and the operation postponed until it can be done with the best technique

The results of surgery are gratifying As previously mentioned, there is usually a prompt reduction in the size of the heart Such favorable results have been obtained in adults as well as in children Nevertheless, when possible it is desirable to operate during childhood Within three months after operation the patient is able to resume full activity It is, however, important to realize that it takes time for the left ventricle to hypertrophy sufficiently to meet the demands normally required by exercise Consequently the full benefit of the operation may not be appreciated for another year During this year there is slow steady

[illegible]

great has been his improvement

PROGNOSIS

The prognosis is far better than is anticipated from the size of the heart Prior to the introduction of antimicrobial therapy and surgery the average length of life was forty years Furthermore, the common causes of death were pneumonia and rheumatic heart disease The recent advances in medicine have therefore greatly improved the prognosis for these patients The original figures included patients with great cardiac enlargement Therefore the life expectancy of a patient with this malformation and a heart of normal size must now be between 70 and 80 years

Cardiac enlargement is an unfavorable sign and is usually considered an indi

cation for operation Surgery has completely altered the prognosis for patients with large auricular septal defects Such defects can be successfully closed and the heart and circulation thereby restored to normal The only major contraindication to surgery is severe pulmonary hypertension Fortunately this complication is rare Therefore the prognosis for the patient with this malformation is generally excellent

SUMMARY

The essential anatomical features of this malformation are a gross defect in the auricular septum, dilatation and hypertrophy of the right auricle and the right ventricle associated with relative enlargement of the pulmonary artery, and a relatively small left ventricle and aorta

The pressure in the left auricle is normally slightly higher than that in the right auricle Hence the shunt is from left to right Anything which increases the pressure in the left auricle, be it rheumatic fever and mitral stenosis or essential hypertension, increases the left to-right shunt The volume of the shunt may be so large that there is excessive circulation to the lungs and the systemic circulation is starved

The malformation is more common in women than in men

The clinical findings in early infancy are those associated with a large left to right shunt namely, failure to gain and repeated pulmonary infections In older patients there may be slight limitation of activity These patients are very susceptible to rheumatic fever but with the decrease in the incidence and severity of rheumatic fever the clinical manifestations of this malformation have changed

The outstanding clinical findings are those of a frail build and left sided chest deformity in a patient with no cyanosis but with a history of repeated pulmonary infections

The outstanding cardiac findings are great enlargement of the right auricle and the right ventricle The pulmonic second sound is accentuated and widely split A systolic thrill may be palpable over the pulmonary area Usually there *is a systolic murmur in the second and third left interspaces* which is readily audible posteriorly in the interscapular region and often there is a mid diastolic murmur at the apex

When in addition there is mitral stenosis, either congenital or acquired, the malformation is known as Lutembacher s syndrome The left auricle is not enlarged by the obstruction at the mitral valve, as the defect in the auricular septum acts as an escape valve Thus the mitral stenosis increases the left to-right shunt

Pulmonary insufficiency and auricular fibrillation are late complications, as is cardiac failure

Fluoroscopic examination shows great enlargement of the right auricle and right ventricle, fullness of the pulmonary conus, and increased pulmonary vascularity Usually there is a hilar dance

The electrocardiogram shows a tendency to right axis deviation and evidence of an incomplete right bundle branch block

The hematology is usually normal

Cardiac catheterization shows that the shunt is at the auricular level The pressure in the right ventricle is elevated but the pressure in the pulmonary artery is usually slightly less than that in the right ventricle

Angiocardiography may show evidence of either a left to-right shunt or, as the injection of dye raises the pressure in the right auricle, there may be a right to-left shunt Usually there is evidence of re-circulation of dye through the right side of the heart

Selective angiocardiography may be useful to differentiate an ostium secundum defect from an ostium primum defect

The condition is to be differentiated from other malformations of the heart, especially anomalies of the pulmonary venous return, defects in other parts of the auricular septum high ventricular septal defects, patency of the ductus arteriosus rheumatic heart disease, primary pulmonary hypertension, idiopathic dilatation of the pulmonary artery, a Taussig Bing malformation combined with an anomaly of the pulmonary venous return, pulmonary tuberculosis, mediastinal tumors and substernal thyroid

The common complications are rheumatic fever, pneumonia and pulmonary infections and cardiac arrhythmias Occasionally there may be symptoms of pressure on the recurrent laryngeal nerve or venous stasis and thrombus formation Subacute bacterial endocarditis seldom occurs with this malformation Pregnancy is remarkably well tolerated

Medical treatment is directed to the prevention of pneumonia and of recurrences of rheumatic fever and the control of cardiac arrhythmias Digitalis is indicated if there is evidence of cardiac failure or cardiac arrhythmias

Surgical closure of an auricular defect restores the heart and circulation to normal The risk of operation is slight Therefore, if there is cardiac enlargement or symptoms of shortness of breath, surgical closure of the defect is indicated

The prognosis is good and may be further improved by medical care and rendered excellent by surgical correction of the condition

C Persistent Ostium Primum

Persistence of the ostium primum represents an earlier arrest in the development of the heart than does a defect of the ostium secundum type It is due to the failure of the first of the primitive openings in the auricles to close

NATURE OF THE MALFORMATION

The defect lies in the lower portion of the auricular septum in the position of the ostium primum The base of the defect is formed by the mitral and tricuspid valves as they arise from the ventricular septum (see Figure XXIII-4) Frequently there is a cleft in the mitral valve which renders that valve insufficient (see Figure XXIII-3) A similar cleft may occur in the tricuspid valve but this is less common When the mitral and tricuspid valves are competent, the malformation is closely similar to a defect which occurs higher up in the auricular septum (see Figure XXIII-1) If, however, as so frequently occurs, there is a cleft in the mitral valve, the malformation produces a distinctive clinical syndrome

COURSE OF THE CIRCULATION

The blood from the right auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs The oxygenated blood is returned to the left auricle, thence some blood is shunted into the right auricle and the remainder flows into the left ventricle Most of the blood in the left ventricle is pumped out through the aorta to the systemic circulation but, inasmuch as there is a cleft in the mitral valve, some blood from the left ventricle is regurgitated into the left auricle and some blood is shunted directly from the left ventricle into the right auricle The blood from the systemic circulation is returned in the normal manner to the right auricle The blood which is shunted into the left auricle still further raises the pressure in the left auricle and this in turn increases the shunt from the left auricle to the right auricle Thus the right auricle not only receives its normal quota of venous blood from the body but also receives some fully oxygenated blood from the left auricle and from the left ventricle Consequently a large volume of oxygenated and venous blood flows from the right auricle to the right ventricle There the cycle starts again (see Diagram XXIII-3)

PHYSIOLOGY OF THE MALFORMATION

The insufficiency of the mitral valve renders this malformation more serious

than other types of auricular defects The mitral insufficiency causes blood to be regurgitated into the left auricle This has a twofold effect It raises the pressure in the left auricle and increases the left to-right shunt, it also decreases the efficiency of the left ventricle and thereby increases the work required of that ventricle to maintain the systemic output Blood is also shunted directly from the left ventricle into the right auricle, as evidenced by a jet of blood which can be felt to come through the defect into the right auricle with each ventricular systole Any increase in the left to-right shunt increases the volume of blood returned to the left auricle Thus the work of both sides of the heart is increased The malformation may cause progressive cardiac enlargement The overwhelming shunt is from left to right, the oxygen saturation of the arterial blood is normal The pressure in the right ventricle is slightly elevated but it does not reach systemic pressure The pressure in the pulmonary artery is directly proportional to or slightly less than that in the right ventricle

CLINICAL FINDINGS

The clinical findings are similar to those produced by other malformations with a large left to-right shunt

Left sided chest deformity is less commonly seen, as the malformation places a strain on both ventricles The chest is usually barrel shaped

Cyanosis is absent, as the predominant shunt is from left to right There is no clubbing of the extremities

Dyspnea is common, especially as this condition leads to progressive cardiac enlargement and chronic cardiac failure

Pulmonary congestion frequently occurs Attacks of bronchitis and pneumonia are common

The liver is often enlarged

Edema is a late manifestation of cardiac failure

CARDIAC FINDINGS

The heart is enlarged both to the right and to the left There is a systolic heave at the apex and a right ventricular lift is palpable over the precordium Over a period of years the malformation leads to slow, progressive cardiac enlargement

The pulmonary second sound is accentuated and reduplicated

A systolic thrill is palpable along the left sternal border

Diagram XXIII–3

Head and upper extremities

R Lung

L Lung

SVC

PA

Pul A

Aorta

PV

Pul V

Pulmonary Art

LA

RA

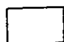
LV

RV

Descending Aorta

IVC

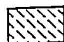
Trunk and lower extremities



Arterial blood (fully saturated)



Venous and arterial blood
Cyanosis visible



Small admixture of venous blood
No visible cyanosis



Venous blood

DIAGRAM XXIII–3

Persistent ostium primum with a cleft in the mitral valve

The essential features of this malformation are a defect in the auricular septum which lies at the base of the auricles and a mitral or tricuspid valve which is cleft and thereby insufficient

The blood in the right auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs, where it is oxygenated and returned by the pulmonary veins in the normal manner to the left auricle Inasmuch as the pressure is higher in the left auricle than in the right auricle some blood is shunted through the auricular defect into the right auricle and the remainder of the blood in the left auricle flows into the left ventricle The insufficiency of the mitral valve means that with each ventricular systole some of the blood from the left ventricle is forced back into the left auricle and some blood is shunted directly from the left ventricle to the right auricle the remainder of the blood in the left ventricle is pumped out through the aorta to the systemic circulation and is returned by the *superior vena cava* and the *inferior vena cava* to the right auricle There it meets the oxygenated blood shunted from the left auricle and the left ventricle into that chamber Consequently the blood which flows into the right ventricle has a relatively high oxygen saturation

Clinical diagnosis The patient shows no cyanosis but may show dyspnea on exertion The heart is slightly enlarged Usually the pulmonic second sound is accentuated and there is a harsh systolic murmur over the *precordium* and a systolic murmur at the apex which is transmitted to the axilla The electrocardiogram shows a left axis deviation and evidence of a right bundle branch block or combined hypertrophy A vectorcardiogram taken in the frontal plane shows that the QRS loop swings in a counterclockwise direction and usually lies above the iso electric point Cardiac catheterization shows that the shunt is at the auricular level

A harsh systolic murmur is maximal over this area This murmur has a harsh, rasping quality similar to that produced by a ventricular septal defect, in addition a *mid diastolic murmur* is audible at or just within the apex When there is *mitral insufficiency,* there is also a *harsh systolic murmur* at the apex which is well *transmitted to the axilla*

Cardiac failure occurs insidiously A gallop rhythm is frequently heard There are rales at the bases of the lungs and engorgement of the liver There may be edema of the extremities

X RAY AND FLUOROSCOPIC FINDINGS

The heart is enlarged to the right and to the left There is fullness of the main pulmonary artery and of its branches The hilar shadows are dense and upon fluoroscopy show conspicuous pulsations (see Figure XXIII–17)

Examination in the left anterior oblique position will show that both ventricles are enlarged Occasionally in the right anterior-oblique position there may be evidence of slight left auricular enlargement, but this is minimal (Figure XXIII–18)



FIGURE XXIII–17 Persistent ostium primum with a cleft in the mitral valve (same patient as in Figure XXIII–3) Child



Left anterior-oblique position



Right anterior oblique position

FIGURE XXIII–18 Persistent ostium primum with a cleft in the mitral valve (same patient as in Figure XXIII–3) Child

ELECTROCARDIOGRAPHIC FINDINGS

The most characteristic finding in a persistent ostium primum with a cleft in the mitral valve is that the standard leads show a left axis deviation and, furthermore, the deflections are often tall and usually show conspicuous notchings, the unipolar precordial leads show evidence either of an incomplete right bundle branch block or of combined hypertrophy (see Figure xxiii–19) Indeed, the finding of a left axis deviation in a patient with an auricular septal defect is strong evidence in favor of a defect of the ostium primum type with a cleft in the mitral valve and with or without a small defect in the ventricular septum

A vectorcardiogram taken in the frontal plane, as shown by Toscano-Barbosa et al,[2] may be extremely helpful in the diagnosis of an ostium primum defect or of a persistent ostium atrioventriculare commune These investigators found that in patients with endocardial cushion defects the QRS loop as it is projected on the frontal plane usually advances in a counterclockwise direction and generally lies mainly superior to the iso-electric point

SPECIAL TESTS

The red blood cell count the amount of available hemoglobin, and *the hematocrit reading* are normal

The oxygen saturation of the arterial blood is normal

Cardiac catheterization will show that there is a marked increase in the oxy

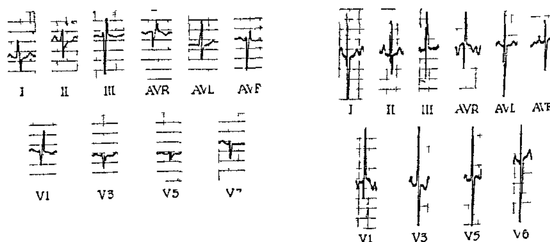


FIGURE XXIII–19 Auricular septal defect of the ostium primum type

Note the left axis deviation and the evidence of combined hypertrophy

gen content of the blood in the right auricle There is sometimes a further increase in the oxygen saturation of the blood in the right ventricle, occasionally the stream line of the shunt is such that the increase in the oxygen content of the blood is detected only in the ventricle The pressures in the right ventricle and the pulmonary artery are usually slightly to moderately elevated Frequently it is possible to pass the catheter into all four chambers of the heart

Angiocardiography is of little value The defect in the auricular septum may or may not be demonstrated Selective angiocardiography, with the placement of the catheter in the left auricle and injection of dye into this chamber, may aid in the determination of whether or not there is any septum at the base of the defect

DIAGNOSIS

The diagnosis is to be entertained in a patient with evidence of a large left to-right shunt who has a harsh systolic murmur similar to that of a ventricular septal defect, x ray evidence of fullness of the pulmonary conus and increased hilar shadows, and electrocardiographic evidence of a left axis deviation and an incomplete right bundle branch block or combined hypertrophy It is substantiated upon cardiac catheterization either by the demonstration that the shunt occurs at the auricular, not the ventricular, level, or by the passage of the catheter into all four chambers of the heart

DIFFERENTIAL DIAGNOSIS

Clinically this malformation resembles a large ventricular septal defect more than any other malformation

A ventricular septal defect differs from this malformation in that the left auricle is frequently enlarged in addition cardiac catheterization shows that there is no increase in the oxygen content of the blood in the right auricle, as compared with that in the superior vena cava and the inferior vena cava, but that there is an increase in the oxygen content of the blood in the right ventricle

TREATMENT

The surgical correction of this malformation is far more difficult than correction of an auricular defect of the ostium secundum type, not only because of the mitral insufficiency, but also because the base of the defect is formed by the mitral and tricuspid valves as they arise from the ventricular septum Consequently there is danger of injury to the valves and injury to the bundle of His

For these reasons the operation is performed with the use of extracorporeal circulation so that the surgeon has sufficient time to correct the valvular abnormality and close the auricular defect Surgical correction is indicated if there is evidence of progressive cardiac enlargement If the patient is asymptomatic, operation may not be indicated because of the increased risk

PROGNOSIS

In the absence of mitral insufficiency the prognosis is as favorable as that of a defect of the ostium secundum type When there is a cleft in the mitral valve, the prognosis is guarded The condition generally leads to progressive cardiac enlargement and death from cardiac failure in childhood or early adult life Surgical correction is more difficult for this defect than for an ostium secundum defect Nevertheless, if there is evidence of cardiac strain, surgical correction is indicated

SUMMARY

A persistent ostium primum represents an early arrest in the development of the heart The resultant defect lies in the lower part of the auricular septum and the base of the defect is formed by the common margin of the mitral and tricuspid valves Frequently there is a cleft in the mitral valve which renders the valve insufficient and thereby produces a distinctive clinical syndrome

The heart is enlarged There is a harsh systolic murmur and a thrill along the left sternal border, and a murmur of mitral insufficiency at the apex Over a period of years there is evidence of progressive cardiac enlargement and cardiac failure

The clinical findings suggest a ventricular septal defect combined with insufficiency of the mitral valve

The electrocardiogram shows a left axis deviation and evidence of an incomplete right bundle branch block or "combined" hypertrophy A frontal vectorcardiogram shows that the excitation wave travels in a counterclockwise direction and the QRS loop usually lies above the iso-electric point

Cardiac catheterization shows that the shunt is at the auricular level

Surgical closure of the defect is more difficult and carries a higher mortality rate than does an operation for a defect of the ostium secundum type

When an ostium primum defect is combined with a cleft in the mitral valve, the prognosis is poor Unless the condition is corrected by surgery, cardiac failure frequently occurs in late childhood or early adult life

D *Persistent Ostium Atrioventriculare Commune*

A persistent ostium atrioventriculare commune represents an even earlier arrest in the development of the heart than does a persistent ostium primum There is a defect in the auricular septum which is continuous with one in the ventricular septum If the defect in the ventricular septum is small, the condition may be closely similar to a defect of the persistent ostium primum type combined with a cleft in the mitral valve Indeed, these two malformations have been classified by Rogers and Edwards [6] as defects of the *atrioventricular canal* Nevertheless, the physiology of the two malformations is different and so are the classic clinical findings, therefore they are discussed as two separate entities

NATURE OF THE MALFORMATION

A persistent ostium atrioventriculare commune occurs when there is an arrest in the development of the heart before either the auricular or the ventricular septa are complete The mitral and tricuspid valves fuse together through the defect as a single atrioventricular valve Frequently, however, the septal leaflets of either or both valves are cleft, thus rendering the valve insufficient (see Figure xxiii-2) The defects in both the auricle and the ventricle are subject to great variation in size Usually the defect in the auricular septum is considerably larger than that in the ventricular septum As mentioned above, when the defect in the ventricular septum is minute, the malformation merges with that of a ostium primum defect with or without a cleft in the mitral valve When the defect in the ventricular septum is large, there may be virtually a single ventricle If the defects in both the auricular septum and the ventricular septum are huge, there is functionally a biloculate heart.

COURSE OF THE CIRCULATION

When the valves are closed there is an opening between the two auricles and one between the two ventricles Furthermore, if one of the valves is insufficient there is a communication between the auricle and ventricle underlying the insufficient valve When the valves are open there is free communication between all four chambers The pressure in the left auricle and the left ventricle is usually higher than that in the right auricle and the right ventricle, and the overwhelming shunt is from left to right

Hence the greater part of the blood in the right auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs, where it is oxygenated The blood from the lungs is returned to the left auricle Inasmuch

as the pressure in the left auricle tends to be higher than that in the right auricle, some blood is shunted from the left auricle to the right auricle The remainder of the blood in the left auricle flows into the left ventricle The pressure in the left ventricle is normally higher than that in the right ventricle, hence some blood is shunted from the left ventricle to the right ventricle The remainder of the blood in the left ventricle is pumped out through the aorta to the systemic circulation and is returned by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again (see Diagram XXIII–4)

PHYSIOLOGY OF THE MALFORMATION

Inasmuch as there is a shunt from left to right both in the auricles and in the ventricles, the work of the right side of the heart is greatly increased As the pressure in the right auricle and the right ventricle rises, some blood may be shunted to the left side of the heart Therefore it is usual to find a large left-to-right shunt and a small right-to-left shunt The condition permits perpetual variation in the volume and the direction of the shunts and leads to progressive cardiac enlargement The right side of the heart carries a far greater load than does the left There is predominant enlargement of the right auricle and the right ventricle As the left to right shunt increases, the work required of the right ventricle increases and the pressure in that chamber rises, if the defect in the ventricular septum is large, the pressure in the left ventricle is transmitted to the right ventricle Indeed, it is usual to find marked elevation of the pressure in the right ventricle to nearly the systemic level, and a proportional elevation of the pressure in the pulmonary artery

CLINICAL FINDINGS

The clinical findings are closely similar to those produced by other defects in the auricular or ventricular septa

Mongolian idiots frequently have a persistent ostium atrioventriculare commune The incidence of this association is so high that, whenever a Mongolian idiot has evidence of a malformation of the heart which causes no visible cyanosis, the possibility should be considered that the defect is of the persistent ostium atrioventriculare commune type The reverse is not true A persistent ostium atrioventriculare commune can occur in an individual with a normal mentality

The appearance of the patient is commonly that of a chronically ill child with a frail build and left sided chest deformity

Cyanosis is usually absent Although there is usually slight oxygen unsatura

tion of the arterial blood, it is seldom sufficient to affect the color of the individual

Clubbing of the extremities virtually never occurs

Cough may be troublesome because of extreme pulmonary congestion

Respirations are usually rapid

Pulmonary infections and *pneumonia* occur quite as commonly in these patients as in patients with other types of auricular septal defects

The liver is frequently enlarged, as chronic cardiac failure is common

CARDIAC FINDINGS

The cardiac findings more closely resemble those of defects in the ventricular septum than those of defects in the auricular septum

The heart is enlarged to the right and to the left The enlargement is mainly caused by the increased work required by the right auricle and the right ventricle

The pulmonic second sound is usually accentuated and reduplicated

A coarse systolic thrill is palpable over the precordium along the left sternal border extending toward the base *The heave* of the right ventricle is also palpable in the same location To those who are trained to feel a lift, the heave almost obliterates the thrill, but to those whose hands are sensitive to vibrations the thrill is more marked than the heave

A murmur is audible when the thrill is palpable, the murmur is *systolic* in time The murmur and the thrill are as harsh and rasping as those produced by a ventricular septal defect In the presence of great cardiac enlargement, it is the rule to hear a systolic murmur over the precordium and both a systolic and a *low pitched mid diastolic murmur* just within the apex A gallop rhythm is common There is nothing which differentiates the apical murmurs from those which occur in any greatly enlarged, poorly functioning heart

Progressive cardiac enlargement is the rule The larger the defect, the more rapid is the rate of enlargement It may, however, require observation over a period of years to demonstrate the progressive increase in the size of the heart

Cardiac failure develops insidiously and becomes chronic Rales in the lungs, engorgement of the liver, and dependent edema are late manifestations

X RAY AND FLUOROSCOPIC FINDINGS

All four chambers are enlarged but the predominant enlargement is of the right auricle and the right ventricle Usually the pulmonary conus is prominent

DIAGRAM XXIII-4

R Lung
L Lung
S.V.C.
P.A.
Aorta
Pul. A.
PV
Pul V
Pulmonary Artery
L.A.
R.A.
L.V.
R.V.
Descending Aorta
I.V.C.
Trunk and lower extremities

Arterial blood (fully saturated)

Venous and arterial blood
Cyanosis visible



Small admixture of venous blood
No visible cyanosis



Venous blood

Diagram XXIII-4

Persistent ostium atrioventriculare commune

The essential feature of this malformation is a defect which involves both the auricular and the ventricular septa. The valves of the mitral and tricuspid orifices fuse together *through the defect*. When the valves are closed there is a defect in the auricular septum and a defect in the ventricular septum. When the valves are open or insufficient, there is free communication between all four chambers of the heart.

The blood from the superior and inferior venae cavae enters the right auricle, flows to the right ventricle, and is pumped out through the pulmonary artery to the lungs. The oxygenated blood from the lungs is returned by the pulmonary veins to the left auricle. Most of the blood from the left auricle flows into the left ventricle. If the pressure in the left auricle is greater than that in the right auricle, some blood will be shunted from the left auricle to the right auricle, pumped around the pulmonary circulation and returned to the left auricle. The greater part of the blood in the left auricle flows to the left ventricle and is pumped out through the aorta to the systemic circulation. Owing to the fact that the pressure in the left ventricle is greater than that in the right ventricle, some blood is shunted from the left ventricle to the right ventricle through the ventricular septal defect. Thus there is a tendency in both the left auricle and the left ventricle for a left-to-right shunt. Just as soon as the pressure in either the right auricle or the right ventricle exceeds the pressure in the left auricle or the left ventricle there will be a reversal in the direction of the shunt. Under such circumstances the shunt will be from right to left. The establishment of such a shunt raises the pressure on the left side of the heart. As soon as the pressure on the left side exceeds that on the right side, the direction of the shunt will again be reversed.

In brief, the mechanism within the heart is such that there is a tendency for a perpetual reversal in the direction of the shunt. This leads to progressive cardiac enlargement. If the defect is small and the volume of the shunt is small, the rate of enlargement is slow. The condition may be compatible with life for a number of years. If the ventricular septal defect is sufficiently large so that the pressure from the left ventricle is transmitted to the right ventricle, the pressure in that chamber and in the pulmonary artery will be raised to the systemic level. For this reason there is usually marked pulmonary hypertension.

Clinical diagnosis. There is enlargement of the right auricle and the right ventricle. In addition there is a systolic murmur and a thrill over the body of the heart and also a low pitched mid-diastolic murmur at the apex. The x ray shows enlargement of all four chambers of the heart and increased hilar markings; fluoroscopy reveals a hilar dance. The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy. A vectorcardiogram taken in the frontal plane shows that the QRS loop swings in a counterclockwise direction and usually lies above the iso-electric point.

The malformation usually causes serious incapacity. When the defect is large, the heart undergoes rapid enlargement and cardiac failure occurs in early childhood.

and the hilar shadows are increased (see Figure XXIII–20) Upon fluoroscopy a hilar dance is frequently seen The left auricle remains of normal size

ELECTROCARDIOGRAPHIC FINDINGS

The standard leads usually show a right axis deviation and the unipolar pre cordial leads show evidence of 'combined' ventricular hypertrophy Heart block occasionally occurs in patients with a persistent atrioventriculare commune The block may be complete or there may be a regular 2 1 or 3 1 heart block The vectorcardiogram shows a similar pattern to that which occurs in a defect of the ostium primum type

SPECIAL TESTS

The red blood cell count, the amount of available hemoglobin and *the hema tocrit reading* are usually a high normal or slightly elevated but extreme poly cythemia does not occur

The oxygen saturation of the arterial blood is usually around 80 per cent

Cardiac catheterization usually shows a marked increase in the oxygen con



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FIGURE XXIII–20 Persistent ostium atrioventriculare commune Child

tent of the blood in the right auricle, as compared with that in the superior vena cava and in the inferior vena cava, and a further increase in the oxygen content of the blood in the right ventricle Unless the ventricular defect is exceptionally small the high pressure in the left ventricle is transmitted to the right ventricle and to the pulmonary artery Furthermore, it may be possible to pass the catheter into all four chambers of the heart

Angiocardiography is of little aid, as the dye is so rapidly dissipated throughout the heart Selective angiocardiography with dye injected into the left ventricle may show the passage of dye to the right ventricle and also to the *left auricle* but this does not prove that there is an auricular defect

DIAGNOSIS

When the patient is a Mongolian idiot who has evidence of a ventricular septal defect and no visible cyanosis, the possibility should be considered that he has a persistent ostium atrioventriculare commune

The condition is to be suspected in an otherwise normal individual when there is evidence of a large left to-right shunt and right sided cardiac enlargement combined with a harsh systolic murmur and slight reduction in the oxygen saturation of the arterial blood The diagnosis is strengthened if there is evidence of progressive cardiac enlargement Cardiac catheterization reveals a shunt at both the auricular and the ventricular level

DIFFERENTIAL DIAGNOSIS

This condition calls for differentiation from other types of auricular defects, especially from a defect of the ostium primum type with a cleft in the mitral valve from anomalies of the pulmonary venous return, from ventricular septal defects and also from a single ventricle and from a biloculate heart

A defect of the ostium primum type combined with a cleft in the mitral valve differs from a persistent ostium atrioventriculare commune in that, in the former, the electrocardiogram shows a left axis deviation and the oxygen saturation of the arterial blood is normal Cardiac catheterization reveals that the pressure in the right ventricle is only slightly elevated, whereas in a persistent ostium atrioventriculare commune the pressure in the right ventricle usually approximates that of the systemic circulation

Anomalies of the pulmonary venous return may be confused with a persistent ostium atrioventriculare commune because both may lead to tremendous cardiac enlargement and severe cardiac failure The murmurs are more variable

and usually less intense in anomalies of the pulmonary venous return than in a persistent ostium atrioventriculare commune Cardiac catheterization usually shows a far greater increase in the oxygen content of the blood in the right auricle in an anomaly of the pulmonary venous return than in the malformation under discussion When there is total anomaly of the pulmonary venous return, the oxygen content of the blood in all four chambers of the heart and in both great vessels is usually the same

A partial anomaly of the pulmonary venous return with an intact auricular septum is to be distinguished from a persistent ostium atrioventriculare commune by the fact that upon cardiac catheterization only the right side of the heart can be catheterized If the auricular septum is intact, it is manifestly impossible to catheterize the left auricle or the left ventricle

A ventricular septal defect, although it may be a serious lesion, seldom causes as great difficulty in early life as does a persistent ostium atrioventriculare commune Frequently the left auricle is slightly enlarged Furthermore, there is no increase in the oxygen content of the blood in the right auricle

A single ventricle in which there is free admixture of venous and oxygenated blood is to be differentiated from a persistent ostium atrioventriculare commune by the finding that the shunt occurs not at the auricular level but at the ventricular level

A biloculate heart in which there is but a single auricle and a single ventricle, may be closely similar to a persistent ostium atrioventriculare commune with a huge defect in the auricles and in the ventricles The finding of the same pressure *in all parts of the common ventricle is evidence of a huge ventricular septal* defect When there is a common ventricle, angiocardiography reveals no difference in the contour of the ventricle in the early and the late films

TREATMENT

Medical treatment is mainly directed toward improvement in compensation and toward the prevention of infections These patients are unusually susceptible to bronchitis and pneumonia, they are also susceptible to acute and subacute bacterial endocarditis It is important to remember that they do not enjoy the freedom from bacterial infection which is common in association with an ostium secundum defect

Surgical correction of this malformation is difficult because of the relation of the defect to the mitral and tricuspid valves and the bundle of His Inasmuch as the defect is low down and involves both the auricular and the ventricular septa,

the use of an extracorporeal circulation is necessary The operation is difficult and the risk is considerably greater than for all other types of auricular defects

Nevertheless, inasmuch as the condition leads to progressive cardiac enlargement and death from cardiac failure, if the heart is greatly enlarged and the compensation is precarious, operation may be necessary

SUMMARY

A persistent ostium atrioventriculare commune is closely related to a defect of the ostium primum type It represents an even earlier developmental arrest than does a persistence of the ostium primum In a persistent ostium atrioventriculare commune, the mitral and tricuspid valves fuse together through the defect

A persistent ostium atrioventriculare commune is relatively common in Mongolian idiots but also occurs in otherwise normal individuals

The predominant shunt is from left to right at the auricular level

The clinical signs are similar to those produced by a large ventricular septal defect

Cyanosis and clubbing are absent

Respiratory infections are common

The heart is enlarged and usually undergoes progressive enlargement Murmurs and thrills resemble those of ventricular septal defects The x ray shows enlargement of all four chambers of the heart with predominant enlargement of the right auricle, the right ventricle, and the pulmonary artery Fluoroscopy generally reveals a hilar dance

The electrocardiogram usually shows a right axis deviation and evidence of combined hypertrophy The vectorcardiogram taken in the frontal plane shows that the QRS loop advances in a counterclockwise direction

The oxygen saturation of the arterial blood in most instances is approximately 80 per cent Cardiac catheterization shows an increase in the oxygen content of the blood in the right auricle and a further increase of that in the right ventricle The right ventricular pressure and the pressure in the pulmonary artery are generally markedly elevated

Angiocardiography is of no great aid in the diagnosis

This malformation requires differentiation from other types of defects in the auricular septum, from anomalies of the pulmonary venous return, and from ventricular septal defects

Surgical closure of a persistent ostium atrioventriculare commune, although

difficult because of the danger of injury to the mitral and tricuspid valves and to the bundle of His, is possible

The prognosis is guarded The malformation leads to progressive cardiac enlargement and death in childhood or early adult life Successful correction of the malformation changes the prognosis from poor to good

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CHAPTER XXIV

VENTRICULAR SEPTAL DEFECTS

DEFECTS in the ventricular septum may be large or small They may occur as an integral part of another malformation or they may constitute the principal or the sole abnormality in the heart It is with such defects that this chapter is concerned Broadly speaking there are two major types—a small ventricular septal defect and a large ventricular septal defect with markedly increased pulmonary blood flow with or without high pressure in the pulmonary artery

A small ventricular septal defect, which is known as the maladie de Roger,[1] is characterized by the single finding of a harsh systolic murmur Nevertheless, it is an error to think that all harsh precordial systolic murmurs are due to ventricular septal defects

Remarkably harsh systolic murmurs may disappear at puberty This phenomenon has led some observers[2] to believe that perforations in the ventricular septum may close at puberty Nevertheless, when one sees an opening in the muscular wall of a ventricular septum smoothly covered with normal endocardium, it is difficult to believe that such an opening could possibly close Ruptures in the ventricular septum are known to occur but the author has never seen a specimen *in which there* was any indication of a previous defect *in the ventricu*lar septum which appeared to have healed spontaneously, nor has she been able to locate such a specimen in the many pathological laboratories available to her Either the defect is there or there is no evidence that one has ever existed Therefore, in the author s opinion, if the murmur disappears, in all probability the original diagnosis was erroneous A ventricular septal defect unless corrected by surgery *persists throughout life*

A large ventricular septal defect differs from a maladie de Roger primarily in size Most large ventricular septal defects lie high up in the membranous portion of the ventricular septum, such defects may or may not be associated with dextroposition of the aorta The size of the defect and the position of the aorta determine the volume of the shunt and the pressure in the pulmonary artery The hemodynamics *are quite different when the volume* of the shunt is large or the pulmonary pressure is elevated Hence these conditions are discussed as separate entities Section A is concerned with the small ventricular septal de

fects, Section B with large defects, and Section C with defects in the ventricular septum which open into the right auricle

A *Maladie de Roger*

NATURE OF THE MALFORMATION

The maladie de Roger was first described by Roger[1] It is the name given to a simple perforation in the septal wall between the two ventricles The location and the size of the defect are subject to great variation

The commonest location of a small ventricular septal defect is relatively high up near the base of the septum, as shown in Figure XXIV–1 The defect may, however, lie low in the ventricular septum, as shown in Figure XXIV–2. Usually there is a single defect and the opening is small, only 0 3 to 0.8 cm in greatest diameter Occasionally there are many small defects in the ventricular septum which lie in the interstices of the muscular wall Under such circumstances the septal wall of the left ventricle is trabeculated but the wall of the right ventricle retains its



FIGURE XXIV–1 Ventricular septal defect (maladie de Roger)





Courtesy of Dr Jesse Edwards and the Mayo Clinic

Right ventricle Left ventricle

FIGURE XXIV–2 Low ventricular septal defect

normal structure (see Figure XXIV–3) Such defects may cause great difficulty (see pages 711 and 715)

COURSE OF THE CIRCULATION

During fetal life a perforation in the ventricular septum causes no significant alteration in the circulation At birth the heart is normal in size The foramen ovale closes normally, the ductus arteriosus undergoes normal obliteration

After birth with the expansion of the lungs, the pressure in the right ventricle falls As soon as there is a significant difference between the pressures in the two ventricles blood is shunted from the left ventricle to the right ventricle If the defect is in the membranous septum, the blood from the left ventricle may be shunted into the outflow tract of the right ventricle and hence into the pulmonary artery

Regardless of the location of the defect, the blood in the right auricle flows into the right ventricle and is pumped out by way of the pulmonary artery to the lungs The oxygenated blood is returned by the pulmonary veins to the left auricle and thence to the left ventricle

Inasmuch as the pressure is higher in the left ventricle than in the right, with each systole a small amount of blood from the left ventricle is shunted through



C i y J Dr J se Ed d d he M yo Cl

Right ventricle



C sy J Dr J Edward d th M y Cl c

Left ventricle

FIGURE XXIV–3 Multiple ventricular septal defects

the defect in the ventricular septum into the right ventricle, or almost directly *into the* pulmonary artery This blood, together with that from the right ventricle, is pumped through the pulmonary artery to the lungs It is returned by the pulmonary veins to the left auricle and thence to the left *ventricle The greater part of the blood in the left ventricle* is pumped out through the aorta to the *systemic circulation* and is returned by the superior vena cava and the inferior vena cava to the right auricle So the cycle continues The course of the circulation is shown in Diagram XXIV-1

PHYSIOLOGY OF THE MALFORMATION

In this malformation the defect is so small that the normal *pressure relations are maintained* Therefore the only effect upon the circulation is the shunting of a small amount of oxygenated blood from the left ventricle to the right ventricle, which re circulates through the lungs

CLINICAL FINDINGS

The condition is usually asymptomatic and may be discovered only upon routine physical examination

Cyanosis never occurs in a maladie de Roger Blood is always shunted from an area of high pressure to one of low pressure, hence it is from the left ventricle to the right ventricle Although on an anatomical basis it is theoretically possible to have a reversal in the direction of the shunt, such a reversal virtually never occurs because the pressure in the right ventricle seldom if ever exceeds that in the left ventricle Even if it did, the size of the defect is usually so small that the volume of the shunt would be insufficient to cause visible cyanosis

Clubbing of the extremities never occurs in a patient with a small ventricular septal defect

Growth and development are usually normal

Exercise tolerance is normal

CARDIAC FINDINGS

The size and the shape of the heart are usually normal If the defect is moderately large, both ventricles may be slightly enlarged and the pulmonary conus dilated

A loud systolic murmur and a thrill in a heart of normal size are the outstanding findings of a ventricular septal defect The murmur is usually audible shortly after birth It has a harsh, rasping quality and is of maximal intensity in

the third and *fourth left interspaces*, close to the sternum The *murmur* is well transmitted to the right and to the left It radiates as does the ripple caused by a pebble thrown into the water The murmur is frequently well heard over the *posterior thorax* but is seldom transmitted beyond the thoracic cage or audible in the vessels of the neck

Although a thrill is not palpable in every case, the defect generally causes a sufficiently great disturbance in the flow of blood to produce a palpable thrill Both the murmur and the thrill are increased in intensity by exercise, the murmur is louder and the thrill is more pronounced in the recumbent than in the erect position

The pulmonic second sound is usually normal, it is never unduly accentuated

A moderately large defect in the ventricular septum may cause cardiac enlargement. The enlargement involves both ventricles and the left auricle The right ventricle undergoes dilatation and hypertrophy because of the large volume of blood shunted through the defect and consequently the right ventricle is required to pump an increased volume of blood into the pulmonary circulation The left auricle and the left ventricle are both enlarged because of the increased volume of blood returned from the lungs to the left side of the heart The right auricle is spared

X RAY AND FLUOROSCOPIC FINDINGS

The x ray findings in the maladie de Roger are significant only in that the heart is normal in size and shape (see Figure XXIV-4) Even in cases with a high ventricular septal defect and slight cardiac enlargement the contour of the heart is essentially normal There may be fullness of the pulmonary conus and increased hilar markings

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram is usually normal and therefore of no diagnostic aid A delay in the intraventricular conduction time is notably *rare* in ventricular septal defects, even when the defect lies at *the base of the* septum in the apparent pathway of the bundle of His

SPECIAL TESTS

The red blood cell count the amount of available hemoglobin, and the hematocrit reading are *normal* The oxygen saturation of the *arterial* blood is normal

Diagram XXIV-1

Head and upper extremities
R Lung
RA
SVC
PV
Aorta
Pul. A
L Lung
Pul V
Pulmonary Art
LA
RA
LV
RV
IVC
Descending Aorta
Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible

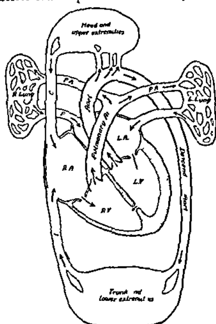

Small admixture of venous blood
No visible cyanosis


Venous blood

Diagram XXIV–1

Ventricular septal defect (maladie de Roger)

In this malformation there is a defect in the ventricular septum Usually the defect is small and lies near the base of the septum as shown in the diagram In a few cases the defect in the septum lies near the apex of the heart as shown in the insert.



Defect in the ventricular septum near the apex of the heart

The blood from the right auricle passes into the right ventricle and is pumped out by way of the pulmonary artery to the lungs The oxygenated blood from the lungs is returned by the pulmonary veins to the left auricle and passes into the left ventricle Most of the blood in the left ventricle is pumped out through the aorta to the systemic circulation and is returned in the normal fashion by the superior and inferior venae cavae to the right auricle Inasmuch as the pressure in the left ventricle is higher than that in the right ventricle, with each systole a small amount of oxygenated blood from the left ventricle is forced through the ventricular septal defect into the right ventricle, where it mixes with the venous blood coming from the right auricle and is pumped out through the pulmonary artery to the lungs All the oxygenated blood from the lungs is returned by the pulmonary veins to the left auricle and flows into the left ventricle So the cycle continues

The defect is usually small hence the volume of the shunt is small and the heart is normal in size and shape Inasmuch as the shunt is from left to right, there is no cyanosis and no clubbing

Clinical diagnosis The outstanding clinical findings are a pronounced systolic thrill and a harsh systolic murmur over the body of the heart The x ray shows a heart of normal size and contour The electrocardiogram is usually normal The patient is asymptomatic

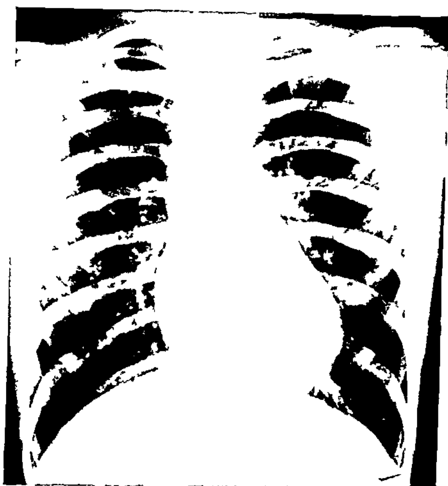


FIGURE XXIV-4 Ventricular septal defect (maladie de Roger) Adult

Angiocardiography is of no diagnostic value

Cardiac catheterization may be necessary to clinch the diagnosis If the catheter is passed from the right auricle into the right ventricle and into the pulmonary artery, blood samples taken from the various chambers will show that the principal increase in the oxygen content of the blood occurs in the right ventricle as compared with that in the right auricle If the defect opens into the right ventricle beneath the pulmonary artery, the principal increase in the oxygen content of the blood may be found in the pulmonary artery The volume of the shunt may be so great as to raise the systolic pressure in the right ventricle The pressure in the pulmonary artery, however, usually remains within normal limits Occasionally the volume of the shunt may be so great that the pulmonary pressure is moderately or greatly elevated (see Section B)

DIAGNOSIS

The diagnosis is based upon the finding of a harsh systolic murmur and a thrill close to the sternum in the third and fourth left interspaces in a person

with a heart of normal size and shape The patient is usually asymptomatic and shows neither cyanosis nor clubbing

DIFFERENTIAL DIAGNOSIS

with other conditions In young infants, both an auricular septal defect and a patent ductus arteriosus may be difficult to differentiate from a defect in the ventricular septum This is especially true if the defect is large In early life, if the growth and development of the individual are normal, there is *no need for* concern

In children a hemic murmur, an abnormally loud functional murmur, or the murmur of aortic stenosis may simulate that of a ventricular septal defect

The murmur which results from anemia may be confused with the murmur of a ventricular septal defect Such a murmur disappears when the hemoglobin rises

Functional murmurs are at times difficult to differentiate from those produced by a ventricular septal defect A loud functional murmur usually has a groaning quality in contrast to the rasping quality of a murmur caused by a ventricular septal defect In addition, a functional murmur is seldom accompanied by a pronounced thrill furthermore, the murmur is rarely transmitted to the back

Aortic stenosis is the commonest malformation to be mistaken for a ventricular septal defect Aortic stenosis, especially in young children, may cause a harsh systolic murmur which is maximal along the left sternal border but not transmitted to the vessels of the neck (see Chapter *xxviii* Section B) The murmur may be even louder than that of a ventricular septal defect but the thrill is usually less pronounced Cardiac catheterization may be necessary to differentiate this murmur from that of a maladie de Roger *Aortic stenosis* is purely a left sided lesion and hence cardiac catheterization will show *no evidence of a shunt*

TREATMENT

A maladie de Roger is usually asymptomatic and requires no treatment If the condition is discovered upon routine physical examination and recognized as such it is wise to advise *the patient* or the parents of the nature of the murmur It should be emphasized that the *malformation* is not serious and *is not likely* to cause any disability There is no need for limitation of activity

Although surgical closure of such a ventricular defect is possible *and* techni

cally not difficult, a small ventricular septal defect has a great tendency to reopen If the sutures pull out, it increases the size of the defect Even if the defect is permanently closed, we do not yet know whether or not the incision of the right ventricle will cause late complications quite as serious as those of the defect itself Therefore as of 1960 operation is seldom indicated

Prophylactic chemotherapy prior to dental extraction, tonsillectomy, etc, is virtually the only treatment that is necessary Even though the defect is small and causes no disability, the condition does render the patient susceptible to subacute bacterial endocarditis No assurance, however, can be given that closure of a ventricular septal defect will be so perfect that the danger of this complication will be eliminated Surgery should not be recommended on this account

COMPLICATIONS

Acute and subacute bacterial endocarditis are the only serious complications The bacteria lodge in the right ventricle at the point where the blood which is shunted through the defect impinges against the wall of the heart Usually the defect is high in the ventricular septum and the blood is driven against the outflow tract of the right ventricle just below the pulmonary orifice There the fungating mass forms In the early stages of the disease, emboli are thrown off into the pulmonary circulation, causing pulmonary infarcts and pneumonic consolidations Positive blood cultures from the systemic circulation may be difficult to obtain As the mass grows, signs of pulmonary stenosis may develop and the murmur at the pulmonary orifice may overshadow that produced by the septal defect

Although subacute bacterial endocarditis is always serious, nevertheless in most instances the infection can be cured by appropriate antimicrobial therapy

PROGNOSIS

The prognosis for a patient with a maladie de Roger is excellent Such persons usually lead long and active lives Even for patients with large ventricular septal defects, so long as the pressure in the pulmonary artery remains normal, the prognosis is reasonably good If, however, the defect is so large as to cause incapacity, the condition can be corrected by surgery (see Section B)

SUMMARY

The maladie de Roger is the outstanding malformation which causes pronounced signs and minimal symptoms The heart is usually normal in size and

... maximal over the body of the heart The ...
shape ...
murmur is ...
There is no clubbing and no cyanosis

The murmur is to be differentiated from both hemic and functional murmurs and also from that of aortic stenosis In infancy it may be difficult to differentiate the malformation from a patent ductus arteriosus or an auricular septal defect In children the murmur may closely simulate that of aortic stenosis In young adults the condition becomes distinctive

The only factor of serious import is the danger of acute and subacute bacterial endocarditis Such infections are always serious, but usually curable with antimicrobial substances

For patients with a maladie de Roger the prognosis is excellent

B *Large Ventricular Septal Defects and the Eisenmenger Complex*

NATURE OF THE MALFORMATION

Large ventricular septal defects may occur anywhere in the ventricular septal wall Most of them occur high up in the membranous septum beneath the aortic valve (see Figure XXIV-5) Many of them are located beneath the tricuspid valve Such defects are occasionally associated with abnormalities in the mitral and/or tricuspid valve (see Figure XXIV-6), if so, they are basically endocardial cushion defects with an intact auricular septum (see Chapter XXIII, Section C) Such a defect involving the ventricular septum and the *tricuspid* valve is shown in Figure XXIV-7, in this instance the *mitral* valve was normal

A high ventricular septal defect which occurs in the membranous septum lies immediately beneath the junction of the right coronary cusp and the septal cusp of the aortic valve Such a defect may or may *not* be associated with slight dextroposition of the aorta and may occasionally be associated with multiple defects (see Figure XXIV-8)

The Eisenmenger complex is a high ventricular septal defect combined with dextroposition of the aorta This malformation has sometimes been considered as a variant of the tetralogy of Fallot *or* a type of partial transposition Actually it resembles the tetralogy of Fallot only in that the aorta may partially over ride the ventricular septum, it differs from the tetralogy of Fallot in that there is no pulmonary stenosis and there is not necessarily right ventricular hypertrophy Furthermore, the hemodynamics of the two malformations are totally different





Courtesy of Dr Jesse Edwards and the Mayo Clinic

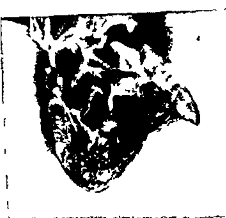
Right ventricle   Left ventricle

FIGURE XXIV–5 High ventricular septal defect

Patients with this malformation do not suffer from inadequate pulmonary blood flow, on the contrary, they suffer from excessive pulmonary blood flow and high pressure in the pulmonary artery

Eisenmenger,[3] in his original description of this malformation in 1897, emphasized the fact that the aorta was slightly dextroposed and consequently arose partially from the right ventricle Under such circumstances the aortic septum cannot meet the ventricular septum, hence there is a high ventricular septal defect (see Figures XXIV–9 and 10) Such defects in the ventricular septum may be large, extending more than 1 0 cm downward in the vertical plane

The degree of over riding of the aorta is subject to considerable variation Usually it is not great, approximately 10 per cent or less Actually surgical experience has shown that in many patients a large ventricular septal defect is not





Court y J Dr J ie Ed d d th M y Cl c

Right ventricle

Left ventricle

Figure xxiv–6 Ventricular septal defect beneath the mitral and tricuspid valves





C t y J D J Ed d d the M y Cl

Right ventricle

Left ventricle

Figure xxiv–7 Ventricular septal defect and cleft tricuspid valve



FIGURE XXIV–8 High ventricular septal defect combined with multiple small ventricular septal defects Left ventricle

C rtesy of D Jes e Ed ds a d the Mayo Cl c

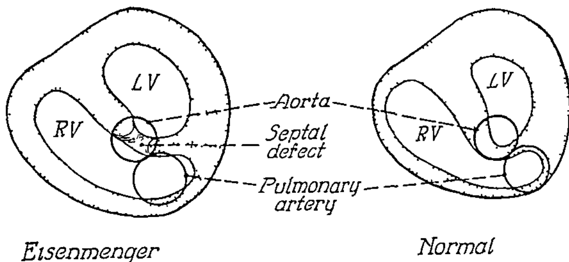


FIGURE XXIV–9 The Eisenmenger complex and normal heart

Cross section near the base of the ventricles showing the relation of the aorta and the pulmonary artery to the ventricular septum



FIGURE XXIV 10 An Eisenmenger complex

The aorta is slightly dextroposed and a high ventricular septal defect lies beneath the non-coronary cusp and the right coronary cusp which is abnormally large and insufficient

associated with any demonstrable over riding of the aorta The question has also been raised whether the abnormal currents of blood could cause the aorta to become progressively further displaced to the right Certain it is that a 10 per cent over riding in a small heart may be so slight as to be difficult to determine, whereas a 10 per cent over riding of the aorta in an adult is perceptible at a glance

In view of the great variation in the extent of the dextroposition of the aorta which is anatomically present and the impossibility of determining this accurately by clinical means it is advisable to regard the Eisenmenger complex as a

syndrome The *Eisenmenger syndrome* may be defined as a heart with a large ventricular septal defect, increased pulmonary blood flow, and high pulmonary pressure The defect usually lies in close proximity to the aortic orifice and the aortic valve is frequently insufficient

The effect of the defect upon the hemodynamics does not depend so much on the location of the defect as upon the force with which the blood is ejected to the lungs The pulmonary vascular bed is so large that it requires an enormous volume of blood to elevate the pulmonary pressure, whereas if the aorta over rides the ventricular septum to such an extent that the right ventricle works against systemic pressure, then regardless of the volume of the shunt, the lungs receive blood under systemic pressure In the author s opinion, the extent of the dextroposition of the aorta is frequently an important factor in both the production of the pulmonary hypertension and the age at which it causes difficulty

COURSE OF THE CIRCULATION

During fetal life a ventricular septal defect, even when there is dextroposition of the aorta, has little if any effect upon the circulation The malformation is readily compatible with fetal life The heart at birth is normal in size

After birth the foramen ovale and the ductus arteriosus undergo normal obliteration The blood from the right auricle flows into the right ventricle The greater part, if not all, of the blood from the right ventricle is pumped out by way of the pulmonary artery to the lungs and is returned by the pulmonary veins to the left auricle and thence to the left ventricle Most of the blood from the left ventricle is pumped out through the aorta to the systemic circulation and returned by the superior vena cava and the inferior vena cava to the right auricle Since the pressure in the lesser circulation tends to be lower than that in the systemic circulation, with each ventricular systole not only is blood pumped into the aorta, but some blood is shunted through the defect into the right ventricle and into the pulmonary artery As long as the pressure in the pulmonary artery is less than that of the aorta, the shunt is entirely from left to right The course of the circulation, as shown in Diagram XXIV-2, is essentially the same as that of the maladie de Roger

If the shunt is huge, or when the aorta over rides the ventricular septum, although most of the blood in the right ventricle is pumped out through the pulmonary artery to the lungs, a small volume of venous blood is pumped out through the aorta to the systemic circulation The blood which is pumped out into the pulmonary artery flows to the lungs, where it is oxygenated, and re

turned in the normal manner to the left auricle, thence it flows to the left ventricle The blood from the left ventricle is pumped out into the aorta and also, through the septal defect, into the pulmonary artery Hence there is both a right-to-left and a left-to-right shunt The volume of the shunt from right to left is frequently so small that there is no visible cyanosis in childhood (see Diagram XXIV-3)

Nevertheless, if the aorta over rides the ventricular septum, the right ventricle pumps against systemic pressure and the pressure in the pulmonary artery is proportionally elevated The high pressure with which the blood is pumped into the lungs leads to injury to the pulmonary vessels and causes progressive narrowing of the pulmonary vascular bed and progressive pulmonary hypertension As the pressure in the pulmonary artery rises, the volume of the left-to-right shunt decreases and sooner or later the pulmonary hypertension, combined with the high pressure in the right ventricle, increases the volume of blood to be shunted from the right ventricle into the aorta Eventually the shunt from right to left becomes of sufficient magnitude to give visible cyanosis (see Diagram XXIV-4)

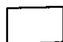
PHYSIOLOGY OF THE MALFORMATION

The essential feature which differentiates both a large ventricular septal defect and an Eisenmenger complex from a maladie de Roger is the high pressure in the right ventricle and in the pulmonary artery The increased pressure in the pulmonary artery is in part due to the tremendous pulmonary blood flow and the compensatory slow opening up of the pulmonary vascular bed, and in part may be due to the over riding of the aorta, which causes the right ventricle to work against systemic pressure The real danger is that the increased force with which the blood is ejected into the lungs will eventually lead to severe intimal changes these changes in turn increase the resistance in the pulmonary vascular bed

The rapidity with which these changes occur is not known Moreover, such changes do not always occur There may be a large pulmonary blood flow with little or no pulmonary hypertension It is important to remember that, because the pressure in the pulmonary artery is abnormally high in early life, it will not inevitably lead to pulmonary hypertension

The author has seen several children in whom, over a period of years, the pulmonary pressure fell In one instance the pulmonary pressure fell from 78/39 at four years of age to 47/22 mm of mercury at nine years of age, in another instance the pulmonary pressure fell from 56/23 to 33/23 between the ages of

DIAGRAM XXIV–2



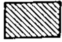
Arterial blood (fully saturated)



Venous and arterial blood
Cyanosis visible



Small admixture of venous blood
No visible cyanosis



Venous blood

DIAGRAM XXIV-2

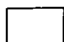
High ventricular septal defect

The essential feature of this malformation is a defect at the base of the aorta which lies in the membranous portion of the ventricular septum Because the defect is so high up, some of the blood from the left ventricle is pumped into the right ventricle and thence into the pulmonary artery

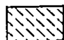
The blood from the right auricle flows into the right ventricle and is pumped out in the normal fa hion through the pulmonary artery to the lungs The blood is re turned by the pulmonary veins to the left auricle and thence it flows to the left ventri cle Most of the blood in the left ventricle is pumped out through the aorta to the sys temic circulation and is returned by the superior and inferior venae cavae to the right auricle Owing to the high ventricular septal defect combined with the high pressure in the left ventricle, some blood from the left ventricle is pumped into the right ventri cle and thence into the pulmonary artery Thereby some oxygenated blood is re-circu lated through the lungs and the volume of blood in the pulmonary circulation is in creased Thus the work required of the heart is increased consequently the heart is slightly enlarged The shunt is from left to right and there is no cyanosis

Clinical diagnosis The patient is asymptomatic and shows no cyanosis The heart is slightly enlarged There is a harsh systolic murmur *over the precordium* the pul monic second sound is normal The x ray shows fullness of the pulmonary conus and increased hilar shadows The electrocardiogram shows a balanced axis and evidence of left ventricular dominance

DIAGRAM XXIV–3


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis

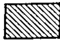

Venous blood

DIAGRAM XXIV-3

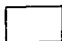
Eisenmenger complex with no visible cyanosis

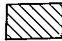
The essential feature of the Eisenmenger complex is the slight dextroposition of the aorta which renders inevitable a high ventricular septal defect and causes the blood to be pumped to the lungs under systemic pressure Therefore there is marked pulmonary hypertension

The blood from the right auricle passes into the right ventricle thence most of the blood is pumped out through the pulmonary artery to the lungs where it is oxygenated The blood from the lungs is returned by the pulmonary veins to the left auricle and thence passes into the left ventricle. From the left ventricle it is pumped out through the aorta to the systemic circulation and is returned in the normal fashion by the superior and inferior venae cavae to the right auricle and thence to the right ventricle Inasmuch as the aorta slightly over rides the right ventricle a small amount of venous blood from the right ventricle is pumped directly into the aorta and passes into the systemic circulation The volume of unoxygenated blood is usually too small to cause visible cyanosis Over a period of years the high ejectile force in the right ventricle causes intimal proliferation in the pulmonary vascular bed which leads to progressive pulmonary hypertension Nevertheless throughout childhood, although arterial oxygen unsaturation gradually increases there is usually no cyanosis

Clinical diagnosis During childhood the patient may be entirely asymptomatic and show no cyanosis The heart is normal in size or slightly enlarged There is a harsh systolic murmur maximal over the precordium and the pulmonic second sound is accentuated *The x ray shows fullness of the pulmonary* conus and increased vascularity The electrocardiogram shows a right axis deviation and evidence of either combined hypertrophy or right ventricular dominance

Diagram XXIV–4


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

Diagram XXIV-4

Eisenmenger complex with cyanosis

This malformation is almost identical with that of the Eisenmenger complex, in which there is no visible cyanosis (see Diagram XXIV-3) Usually the heart is but slightly enlarged The aorta is dextroposed and over rides the ventricular septum, it follows that there is a high ventricular septal defect and pulmonary hypertension

The blood from the right auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs where it is oxygenated, and returned by the pulmonary veins to the left auricle Thence the blood flows into the left ventricle and is pumped out through the aorta to the systemic circulation Inasmuch as the aorta partially arises from the right ventricle some venous blood is pumped from the right ventricle into the aorta Thus the aorta receives a mixture of oxygenated blood from the left ventricle and venous blood from the right ventricle. The blood which is pumped into the aorta flows to the systemic circulation and is returned by the superior vena cava and the inferior vena cava to the right auricle Thence it again flows to the right ventricle So the cycle continues

The high ejectile force in the right ventricle sooner or later causes intimal prolifera tion in the pulmonary vascular bed The intimal proliferation causes progressive nar rowing of the small muscular arteries and leads to progressive pulmonary hyperten sion As the pulmonary pressure rises more and more blood is shunted from the right ventricle into the aorta and eventually the volume of blood so shunted becomes suffi cient to give visible cyanosis

Clinical diagnosis The insidious development of cyanosis at or about the time of puberty is characteristic of this malformation At this time the patient begins to suffer from shortness of breath and may suffer from hemoptyses Dyspnea and cyanosis be come progressively more marked The size of heart remains essentially unchanged but the systolic murmur may become less intense and the pulmonic second sound be comes loud and banging The electrocardiogram shows evidence of increasing right ventricular hypertrophy

four and seven A number of other investigators have reported similar cases The fact that pressure may continue to fall until the age of seven or nine years indicates that the expansion of the lungs may occur even more gradually in a patient with a large ventricular septal defect than in a patient with a large ductus arteriosus

Gasul et al [4] have studied several infants with large left to-right shunts at the ventricular level and high pulmonary pressure who developed pulmonary stenosis with a decrease in the left to-right shunt and in some instances with the development of a right to-left shunt Gasul's cases were proven by catheterization studies The author[illegible] has reported the same phenomenon but her cases were not substantiated by catheterization

In brief, the initial difficulty caused by a large ventricular septal defect is the excessive pulmonary blood flow which is usually associated with high pulmonary pressure The ultimate difficulty depends upon the cause of the high pulmonary pressure If the pulmonary hypertension is due to the slow opening up of the pulmonary vascular bed, the pressure in the lesser circulation may gradually fall *Under such circumstances the patient ultimately shows the clinical syndrome of a large ventricular septal defect with low pulmonary pressure*

Indeed, in some instances it seems probable that the defect does not increase in size with the growth of the heart and consequently the defect becomes relatively smaller Under such circumstances the volume of the shunt decreases, the lungs gradually expand, and the pulmonary pressure falls

In other instances, the increased work of the right ventricle may lead to hypertrophy of the right ventricle and of the crista supraventricularis which in turn leads to the development of infundibular stenosis Thereby the basic malformation is changed from an Eisenmenger complex to a tetralogy of Fallot with or without visible cyanosis

In brief, there are a number of types of ventricular septal defects in which the initial high pressure in the lesser circulation is due to the delay in the normal expansion of the lungs, secondary to a tremendous pulmonary blood flow, and over a period of years the pulmonary pressure may gradually fall It is, however, important to appreciate that the reverse seldom occurs rarely, if ever, does a malformation in which there is excessive pulmonary blood flow and a low pulmonary pressure lead to pulmonary hypertension in childhood or early adult life

The least favorable sequence of events occurs when blood is ejected into the lungs under systemic pressure as in Eisenmenger s original report The contin

ued b[illegible] [illegible] intimal proliferation These changes cause [illegible]
pressure which normally occurs with the gradual [illegible]
sequently over a period of years the vascular resistance *in the lungs increases* *Although* the pulmonary pressure can never be significantly greater than the systemic pressure, the damage to the lungs increases, the pulmonary vascular bed becomes further constricted, and the pulmonary resistance increases Less blood is shunted from the aorta to the pulmonary artery and more blood is shunted from the right ventricle into the aorta The septal defect thus acts as an escape valve and prevents the pulmonary pressure from ever exceeding systemic pressure regardless *of the severity* of the intimal changes Nevertheless, the defect permits the establishment of a right to-left shunt which leads to the late development of cyanosis Such is the sequence of events in the true Eisenmenger complex in which the aorta over rides the ventricular septum and blood is ejected into the pulmonary circulation under systemic pressure

CLINICAL FINDINGS

The primary difficulty during infancy is caused by the excessive pulmonary blood flow

Repeated pulmonary infections menace these infants' lives Frequently a baby scarcely recovers from one infection before he develops another

Atelectasis notably of the right upper lobe, is a common complication Occasionally the heart is so large and the pulmonary pressure so high that it causes collapse of the left lung, especially of the left lower lobe

Sweating may be a troublesome feature Its cause is not clear but it is of common occurrence and may be excessive

A slow weight gain and *a frail body build* are common in infants and children with a huge left to-right shunt

Stunting of growth may be extreme It is usually due to the excessive shunting of blood away from the systemic circulation Extreme stunting of growth should suggest the possibility of hypoplasia of the aorta

Angina and *signs of coronary insufficiency* may occur due to the enormous left to-right shunt The author has seen two such patients One was an infant who was brought to the clinic at the age of four months with a clinical syndrome similar to that produced by the anomalous origin of the left descending coronary artery from the pulmonary artery The other was a child who was repeatedly admitted to the hospital with severe anginal pain associated with

changes in the T waves characteristic of acute coronary insufficiency In this stance the pulmonary pressure was only 40 mm of mercury, thus permitting enormous volume of blood to be shunted from the systemic circulation to lungs It is probably significant that these acute attacks were of relatively sh duration and were followed by prompt improvement in the electrocardiogr The difficulty completely disappeared after closure of the ventricular septal fect

Angina may also occur when pulmonary hypertension is severe Under su circumstances there is usually only a relatively small left to-right shunt, major shunt is from right to left Occasionally children with a ventricular sep defect and high pulmonary pressure suffer from episodes of *syncope*

Numbness and tingling of the extremities may also be a troublesome featu in a patient with a large pulmonary blood flow and a low systemic flow

Dyspnea may or may not occur Infants with this malformation suffer fro pulmonary congestion and excessive pulmonary blood flow Respirations a rapid but attacks of paroxysmal dyspnea do not occur

Clinical improvement almost always occurs if the patient survives infancy he grows to childhood, pulmonary congestion decreases and pulmonary infe tions become less frequent The child gains weight and his exercise toleran improves

A variety of factors may contribute to this improvement In some instanc the pulmonary pressure and the right ventricular pressure drop In such i stances it seems as if the pulmonary vascular bed had expanded slowly an finally was able to adjust to the increased pulmonary blood flow It is also pos ble that, as the heart grows, the ventricular defect does not increase in size an consequently becomes proportionally smaller In other instances, as in those r ported by Gasul, the patient apparently develops infundibular stenosis whic may or may not be sufficient to cause a right to-left shunt but is sufficient t break the high ejectile force of the right ventricle with a resultant fall in th pulmonary pressure In still other instances, the decrease in pulmonary conge tion is due to the development of irreversible pulmonary vascular changes suc as those described by Edwards,[6] and Dammann and Ferencz It is the last men tioned sequence of events which is the most serious

Cyanosis is absent in infancy and is usually not apparent in early childhooc Cyanosis which becomes apparent at or about the time of puberty is character istic of the Eisenmenger complex Although in infancy and early childhood th principal shunt is from left to right, the long continued high pressure in th pulmonary artery leads to ever increasing constriction of the pulmonary vasc

bed As the resistance in the pulmonary vascular bed increases, the pulmonary pressure rises and eventually blood is shunted from the right ventricle into the aorta When the volume of venous blood so shunted becomes sufficiently great, the patient develops cyanosis All present evidence indicates that it is the ever increasing right to-left shunt which leads to the development of cyanosis at or about the time of puberty

Polycythemia develops insidiously as the oxygen saturation of the arterial blood becomes lower In infants and young children the red blood cell count, the amount of available hemoglobin, and the hematocrit reading are usually not increased, but remain at high normal levels Actual increase in the red blood cell count usually becomes evident between ten and fourteen years of age There after the red blood cell count, the amount of available hemoglobin, and the hem atocrit reading steadily rise Eventually the polycythemia may become as great as in other malformations causing persistent cyanosis

Clubbing of the fingers and the toes is relatively slight Inasmuch as clubbing develops secondary to anoxemia and polycythemia, it does not become evident until after the development of cyanosis Thereafter the duration of life may be so short that clubbing may never become pronounced

The exercise tolerance is variable Some children with a large left to-right

slight exertion and at

Most children know

their own exercise tolerance

A patient with a typical Eisenmenger complex may have excellent exercise tolerance during childhood and he does not squat when tired Nevertheless, as puberty approaches he may develop dyspnea Often it is first noticed as the child walks uphill against the wind Although exercise increases cyanosis, the exercise tolerance of individuals with an Eisenmenger complex is always markedly better than that of patients with pulmonary stenosis

The effect of exercise upon the arterial oxygen saturation may be of diag nostic aid Exercise usually produces a slight lowering of the oxygen saturation of the arterial blood but does not cause a rapid drop in the arterial oxygen satura tion such as occurs in a tetralogy of Fallot

Although the resistance in the pulmonary vascular bed is increased, there is no obstruction at the pulmonary orifice, consequently the volume of the pul monary blood flow can be increased with exercise Therefore, as Bing et al [8] have shown, exercise frequently causes a rise in the oxygen consumption per liter of ventilation and a fall in the oxygen saturation of the arterial blood

Hemoptyses may occur in adult life The rupture of the pulmonary vessels

is probably due to the high pressure under which the blood is ejected to the lungs

Hoarseness and *a brassy cough* are late complications Both are probably due to the pressure upon the recurrent laryngeal nerve exerted by the huge pulmonary artery The mechanism is similar to that which may occur in an atrial septal defect[9]

CARDIAC FINDINGS

The heart is usually slightly to moderately enlarged The malformation increases the work required of both ventricles The right ventricle always has an increased volume of blood to pump, furthermore, if there is dextroposition of the aorta, the right ventricle works against systemic pressure and also against the increased resistance in the pulmonary vascular bed The work of the left ventricle is increased in infancy and childhood by the large left to-right shunt as the excessive volume of blood directed to the lungs is returned to the left auricle and the left ventricle Hence the left auricle may be enlarged and so may the left ventricle No strain, however, is placed on the right auricle, this chamber is usually of normal size Consequently the enlargement of the heart lies mainly to the left of the sternum It is important to appreciate that this malformation usually does not lead to progressive cardiac enlargement

The pulmonic second sound varies in intensity and thereby gives an important clue to the height of the pulmonary pressure As the pressure in the pulmonary artery rises, the second sound over the pulmonary area increases in intensity, it is often reduplicated but not widely split Under such circumstances the closure of the pulmonic valve becomes readily palpable

A systolic murmur and a thrill maximal along the left sternal border at the base of the heart are of common occurrence The intensity of the murmur is variable Broadly speaking, the murmur is extremely harsh and rasping when there is a marked gradient in pressure between the two ventricles For this reason the murmur may decrease in intensity as pulmonary hypertension develops Indeed, the author has seen two patients with proven Eisenmenger complex in whom no murmur was audible

Murmurs and thrills are, however, not solely dependent upon the relative pressure in the two chambers but are also produced by the crossing of the two circulations In an Eisenmenger complex there is a left to-right and also a right to-left shunt and there is usually a murmur In contrast to this, in a tetralogy of Fallot with extreme pulmonary stenosis or pulmonary atresia, when the shunt

is only from the right ventricle into the aorta, a similar over riding of the aorta does not lead to the production of a murmur

A mid-diastolic rumble best heard just within the apex, is extremely common in patients with cardiac enlargement and a poorly functioning heart

An early decrescendo diastolic murmur may be audible along the left sternal border This murmur may be caused by pulmonary insufficiency associated with the enormous dilatation of the pulmonary artery and the pulmonary hypertension or by aortic insufficiency The etiology of the aortic insufficiency is not known The aortic insufficiency may be the result of an alteration in the pressure caused by the unequal size of three aortic cusps, or it may be secondary to acquired disease Usually there is no history of a previous rheumatic infection or of a prolonged illness which might be interpreted as bacterial endocarditis The outstanding feature which differentiates aortic insufficiency from pulmonary insufficiency is the occurrence of pronounced peripheral signs of aortic insufficiency

The coronary orifices are frequently displaced upward to such an extent that the efficiency of the coronary circulation is impaired Although, as previously mentioned, the symptoms of angina may in rare instances occur in infancy and early childhood, the condition usually causes no difficulty until in older individuals it produces or aggravates the symptoms of coronary insufficiency

Mitral valvular abnormalities either insufficiency or stenosis increase the strain on the left auricle Either of these abnormalities when severe will cause back pressure on the lungs, which in turn will cause pulmonary hypertension Each of these lesions produces its characteristic murmur at the apex

Cardiac arrhythmias may occur Extrasystoles are a common complaint Patients with large ventricular septal defects may suffer from attacks of paroxysmal tachycardia

X RAY AND FLUOROSCOPIC FINDINGS

There are two distinct contours of the heart which may occur in this malformation The difference in the contour depends upon the size of the pulmonary artery

When the pulmonary artery is of normal size, the heart may have a boot shaped appearance as in Figures XXIV–11 and 12 Usually, however, the pulmonary artery is enlarged and the contour of the heart, except for the relatively small size of the right auricle approaches that of an auricular septal defect, as in Figure XXIV–13 In the anterior posterior position there is definite fullness of



Before operation



After operation

FIGURE XXIV-11 Ventricular septal defect Infant

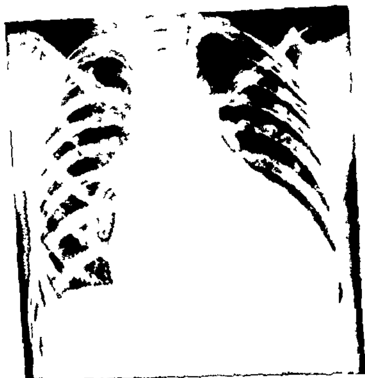
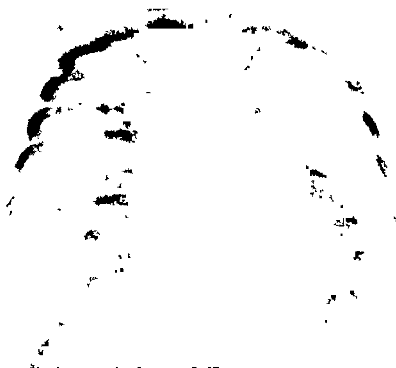


FIGURE XXIV–12 An Eisenmenger complex with a pulmonary artery of normal size Child

the pulmonary conus and of the pulmonary artery Furthermore, the shadows at the hili of the lungs are conspicuous and pulsations in the hilar vessels are readily visible In the left anterior-oblique position both ventricles are seen to be slightly enlarged In the right anterior oblique position the esophagram generally shows evidence of left auricular enlargement and also offers confirmatory evidence of the large size of the pulmonary artery As the child grows the diaphragm drops down, consequently in the adult there may be fullness of the pulmonary conus and minimal cardiac enlargement (see Figure XXIV–14)

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiographic findings vary with the amount of work required of the right ventricle To a large extent the electrocardiogram reflects the pressure in the pulmonary artery When there is increased pulmonary blood flow but pulmonary pressure is low, the main strain falls on the left ventricle as it is required to pump the increased volume of blood which has circulated through the lungs Under such circumstances the standard leads may show a balanced



Before operation



After operation

FIGURE XXIV–11 Ventricular septal defect Infant

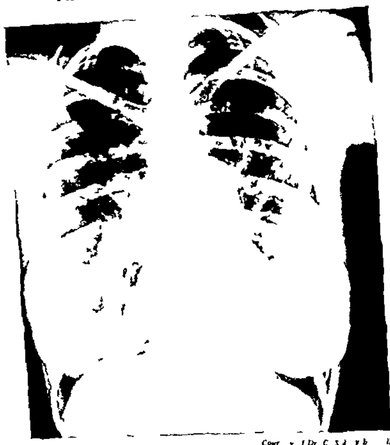
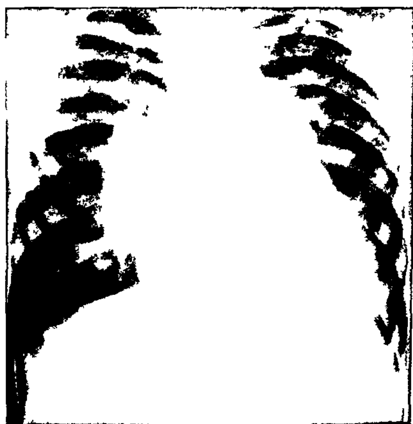


FIGURE XXIV–14 An Eisenmenger complex with a large pulmonary artery Adult

axis or even a left axis deviation and the unipolar precordial leads show evidence of left ventricular dominance (see Figure XXIV–15) When there is marked pulmonary hypertension and the pressure in the right ventricle is correspondingly high, the standard leads show a right axis deviation and the unipolar precordial leads show evidence of right ventricular hypertrophy (see Figure XXIV–16) Often there is moderate pulmonary hypertension combined with an increased circulation to the lungs and the precordial leads show evidence of combined hypertrophy in that there is evidence of right ventricular hypertrophy in V_1 and left ventricular dominance in V_6 (see Figure XXIV–17) In the evaluation of the electrocardiogram due consideration must be given to the age of the patient, because as the child grows the pattern of a right axis deviation combined with balanced precordial leads gradually changes, so that by the time he reaches maturity the electrocardiogram shows a balanced axis and left ventricular domi



Before operation



After operation

FIGURE XXIV–13 Ventricular septal defect Child

nance Nevertheless, careful evaluation of the electrocardiogram is a great help in the estimation of the pressure in the pulmonary artery and hence in the evaluation of the surgical risk (see below)

SPECIAL TESTS

Determination of the circulation time may be of help When there is a right to-left shunt from the pulmonary artery into the aorta, the circulation time becomes abnormally short Therefore this simple test may suffice to demonstrate the over riding of the aorta in an Eisenmenger complex

Analysis of the oxygen saturation of the arterial blood is of great importance In infants and young children the oxygen saturation of the arterial blood is usually normal in older children and young adults, as a right to-left shunt develops, there is unsaturation of the arterial blood before the shunt is sufficiently large to cause visible cyanosis It is however, extremely important to diagnose the condition before the development of cyanosis Therefore cardiac catheterization is often advisable

Cardiac catheterization is of diagnostic value only provided the pulmonary artery is entered and both blood samples and pressure readings are obtained from the various chambers and great vessels Determination of the oxygen content of the blood taken from the various chambers will show an increase in the oxygen content of the blood in the right ventricle as compared with that in the right auricle, usually there is a further increase in the oxygen content of the blood in the pulmonary artery There may or may not be a reduction in the oxygen saturation of the arterial blood Only rarely is it possible to catheterize the aorta When the systolic pressure in the right ventricle is elevated, the systolic pressure in the pulmonary artery is correspondingly high and usually the diastolic pressure in the pulmonary artery is also elevated

Repeated catheterization of patients of various ages or of the same patient at different times may show variation in the pulmonary pressure It is important to appreciate that if the pulmonary pressure is not elevated, the overwhelming probability is that the pressure will remain normal for many years, and probably throughout the life of the individual Indeed, there is no evidence that a patient with a ventricular septal defect and a normal pulmonary pressure is in the slightest danger of developing pulmonary hypertension in adult life

In the Eisenmenger complex the pulmonary pressure is always abnormally high Furthermore there is progressive injury of the pulmonary vascular bed and consequently progressive increase in the pulmonary vascular resistance The

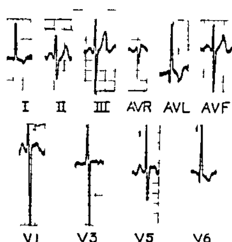


Figure xxiv–15 Ventricular septal defect with low pulmonary pressure

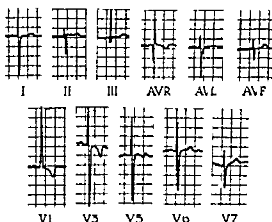


Figure xxiv–16 Ventricular septal defect with high pulmonary pressure (Eisenmenger complex)

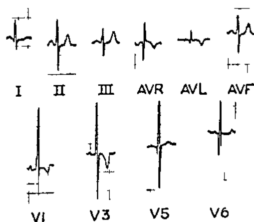


Figure xxiv–17 Ventricular septal defect with pulmonary hypertension and hypertrophy of both ventricles

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SPECIAL TESTS

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In the Eisenmenger complex the pulmonary pressure is always abnormally high Furthermore there is progressive injury of the pulmonary vascular bed and consequently progressive increase in the pulmonary vascular resistance The

ventricular septal defect, however, acts as an escape valve and thereby prevents the pulmonary pressure from ever rising significantly above the systemic pressure even though there is continued narrowing of the pulmonary vascular bed In older patients the pressure in the two circulations is approximately the same and there is usually evidence of a right to-left shunt as well as a left to-right shunt

When there is pulmonary hypertension, it is important to determine whether or not the pulmonary hypertension is reversible The effect of the inhalation of oxygen gives additional valuable information [10] If the pulmonary blood flow increases or the pulmonary pressure decreases, it indicates that the pulmonary resistance is not "fixed"

Angiocardiography may be of aid in the visualization of both the size and the position of the aorta, and also in the demonstration of additional anomalies

The size of the aorta is of importance if operation is contemplated, because if the aorta is hypoplastic it may be unable to carry the full cardiac output Indeed, if the aorta is extremely small, operation is contraindicated Severe hypoplasia of the aorta usually causes stunting of growth, for this reason, the physical development of the patient offers a clue as to its size Therefore, when the patient is markedly underdeveloped, angiocardiography is usually indicated

Dextroposition of the aorta permits simultaneous opacification of the aorta and the pulmonary artery, which is best seen in the anterior posterior view It is a curious fact that in some instances early opacification of the aorta is visible in the anterior posterior series of films and is not found in the lateral series Such a finding in no way invalidates the evidence of dextroposition of the aorta Nevertheless, early visualization of the aorta in the anterior posterior film is seldom evident until after the establishment of the right to-left shunt

When a ventricular septal defect is complicated by mitral stenosis or mitral insufficiency, the dye lingers in the huge left auricle for a long time

DIAGNOSIS

The diagnosis of a large ventricular septal defect is based on the finding of a harsh systolic murmur and x ray evidence of a large left to-right shunt in a patient with no cyanosis Infants usually show a slow weight gain, have a frail body build, and suffer from repeated attacks of pneumonia or atelectasis

In infancy and childhood, in addition to the slight to moderate cardiac enlargement and the harsh systolic murmur, the pulmonic second sound may or may not be accentuated If the second sound over the pulmonary area is not ac

centuated and the electrocardiogram shows evidence of left ventricular dominance, the probability is that the pulmonary pressure is but slightly elevated If, on the other hand, the second sound over the pulmonary area is markedly accentuated and the electrocardiogram shows evidence of right ventricular hypertrophy, the ventricular septal defect is probably complicated by high pulmonary pressure that is, the malformation is an Eisenmenger complex

The insidious development of cyanosis at or about the time of puberty is characteristic of the Eisenmenger complex Whenever possible, diagnosis of this malformation should be made in early childhood, because it should be corrected before there is serious injury to the pulmonary vascular bed

DIFFERENTIAL DIAGNOSIS

In infants and young children prior to the development of cyanosis, a large ventricular septal defect must be differentiated from other malformations with increased pulmonary blood flow, especially a large patent ductus arteriosus before the development of a continuous murmur, an aortic septal defect, anomalies of the pulmonary venous return, auricular septal defects of both the ostium secundum and the ostium primum type and also a persistent ostium atrioventricu lare commune In older children and young adults the malformation may require differentiation from primary pulmonary hypertension, from an auricular septal defect combined with pulmonary hypertension, from pure pulmonary stenosis, and occasionally from a tetralogy of Fallot

A large patent ductus arteriosus or *an aortic septal defect* may cause great cardiac enlargement with only a systolic murmur because of the high pulmonary pressure Under such circumstances an aortogram may be necessary to differentiate these two malformations from a large ventricular septal defect

Anomalies of the pulmonary venous return may occasionally require differentiation from ventricular septal defects The murmurs are variable and may occasionally be as harsh as those of a ventricular defect. Cardiac catheterization will show that the increase in the oxygen content of the blood occurs in the auricle In doubtful cases angiocardiography may be of additional aid in the differentiation of the two conditions

An auricular septal defect differs from a ventricular septal defect in that the systolic murmur is less intense and frequently better heard in the interscapular region than over the precordium, furthermore, the pulmonic second sound is not as loud as is common in ventricular defects and is more widely split Cardiac catheterization shows an increase in the oxygen content of the blood in the right

auricle in comparison with that in the superior vena cava and in the inferior vena cava

An auricular septal defect combined with high pulmonary pressure and a right to left shunt may closely simulate a heart of the Eisenmenger type This combination of anomalies is usually associated with great dilatation of the right auricle, which is rare in the malformation under discussion Cardiac catheteriza tion may be necessary to make a definite diagnosis

An ostium primum defect with a cleft in the mitral valve may show clinical findings closely similar to those in a large ventricular defect Usually there is an accentuated second sound over the pulmonary area and a harsh systolic murmur and a thrill which are maximal close to the sternum in the third and fourth left interspaces The electrocardiogram shows a left axis deviation and evidence of either a right focal bundle branch block or of "combined" ventricular hyper trophy The oxygen saturation of the arterial blood is normal The pressure in the right ventricle may be slightly elevated but seldom, if ever, reaches the level of the systemic pressure

A persistent ostium atrioventriculare commune is similar to an ostium primum defect except that the defect extends into the ventricle Consequently, with a large ventricular defect, the pressure in the right ventricle approaches that of the left ventricle and there is often a further increase in the oxygen content of the blood in that chamber

In both of the above mentioned conditions it is common for the catheter to slip into the left auricle and also into the left ventricle, whereas unless a special effort is made it is virtually impossible to catheterize the left ventricle through a high ventricular septal defect

A maladie de Roger or *a small high ventricular septal defect* differs from a large ventricular septal defect in that the heart is usually of normal size and the pulmonic second sound is not unduly accentuated The pressure in the right ventricle and the pressure in the pulmonary artery are within normal limits or but slightly elevated

Primary pulmonary hypertension is characterized by a marked accentuation of the pulmonic second sound The systolic murmur is seldom loud and may be absent Evidence of pronounced pulmonary insufficiency is more frequently associated with primary pulmonary hypertension than with an Eisenmenger complex Cardiac catheterization confirms the diagnosis of pulmonary hyper tension and shows no change in the oxygen content of the blood throughout the right side of the heart

"Pure" pulmonary stenosis is seldom confused with a ventricular septal de

fect, as the murmur is maximal over the pulmonary area and the second sound at the base is markedly diminished or totally absent

A tetralogy of Fallot is confused with an Eisenmenger complex only in those cases in which there is fullness of the pulmonary conus and slight pulsation of the hilar shadows Patients with a tetralogy of Fallot are usually more limited in their exercise tolerance than are those with an Eisenmenger complex Almost always there is a history of attacks of paroxysmal dyspnea or of squatting in early childhood Cardiac catheterization and the measurement of the pressure in the pulmonary artery readily differentiate the two malformations

ADDITIONAL CARDIAC ABNORMALITIES

Multiple ventricular defects may occur in combination with a high ventricular septal defect. Such defects may be extremely difficult to see, as they lie buried in the trabeculation of the septal wall, as shown in Figure XXIV-8 Although such defects may appear to be insignificant in size, they may be so numerous as to cause a large left to-right shunt Such defects often cause marked elevation of the pressure in the right ventricle and in the pulmonary artery

Aortic insufficiency is relatively common In the Eisenmenger complex not infrequently in addition to the abnormal position of the aorta, there is an abnormality of the aortic valve The right coronary cusp which lies above the septal defect is often abnormally large and deep and is sometimes situated at a lower level than are the other two cusps, as shown in Figure XXIV-10 The aortic valve may become insufficient Whether this is due to the abnormal currents of blood or to the abnormal distribution of the pressure upon the cusps, is not known Nevertheless this is the cusp which usually becomes insufficient The coronary orifices are often displaced upward and are given off from the aorta at or above the top of the semilunar cusps The pulmonary artery occupies its normal position and is of normal size or slightly enlarged

Hypoplasia of the aorta may occur The pulmonary artery is normally slightly larger than the aorta In Eisenmenger s original case the pulmonary artery was abnormally large but the aorta was not severely hypoplastic Occasionally, however, the aorta may be abnormally small, indeed, so small that it is unable to carry the entire output of the left ventricle

Anomalies of the mitral valve may also occur in combination with a large ventricular septal defect There may be either congenital mitral insufficiency or stenosis Either of these conditions mav lead to great left auricular enlargement and either in itself may cause pulmonary hypertension

Pulmonary stenosis may occur in combination with a ventricular defect and

a large left-to-right shunt In such instances the pulmonary stenosis may be sufficient to break the force with which the blood is ejected into the pulmonary artery but insufficient to cause a right to left shunt

TREATMENT

Lillihei[11] was the first to demonstrate that it is possible to close a ventricular septal defect under direct vision This means open heart surgery and the use of a pump and oxygenator The operation is long and difficult and should not be lightly undertaken Nevertheless it can be done with extraordinary success For many patients, especially those with a large ventricular septal defect and abnormally high pulmonary pressure, closure of the defect restores the heart and circulation to normal, furthermore, the operation prevents the development of irreparable damage to the lungs The mortality rate is relatively low when the pulmonary pressure is appreciably lower than the systemic pressure Indeed, if the pulmonary pressure is but 40 per cent of the systemic pressure, the mortality rate, in experienced hands, is remarkably low When the pulmonary pressure is approximately 70 per cent of the systemic pressure, the mortality rate is considerably higher Any reversal of the shunt means that there is severe pulmonary hypertension Under such circumstances the mortality rate is extremely high

Open heart surgery requires an experienced team not only of surgeons but also of anesthesiologists, physiologists, and clinicians Careful attention must be paid to the blood matching, the blood volume, the clotting mechanism, the blood chemistries, and the electrolyte balance Recent studies at the Mayo Clinic[1] show that postoperative limitation of fluids is extremely important

This book is primarily for clinicians, not surgeons it is concerned with the clinical diagnosis, the medical treatment, and the indications for surgery, but not with the surgical technique or with anesthesia For the technique of open heart surgery and the immediate postoperative care and complications, the reader must consult the appropriate authorities The clinical diagnosis and the selection of patients for operation and the resumption of activity after operation are the responsibility of the clinician

Many of these patients suffer from chronic heart failure and repeated respiratory infection It is the clinician s responsibility to see that the patient is in the best possible condition for operation The use of digitalis in open heart operation is controversial In some instances, digitalis seems to increase the liability to arrhythmias, therefore it should be used with caution Some surgeons prefer to have the patient off digitalis before operation If such is the plan, digoxin may be

of great help *The prompt* excretion of this preparation enables the patient to receive the *full benefit of* digitalis until a couple of days prior to operation Immediately after operation many physicians give digitalis because of the incidence of cardiac failure in the early postoperative period

INDICATIONS FOR OPERATION

The *principal indication* for operation is cardiac enlargement *with increased* pulmonary blood flow and increased pulmonary pressure, especially when combined with incipient or chronic cardiac failure

Although *during* the first year or two of life the child may suffer from *repeated respiratory infections*, pneumonia, and chronic cardiac failure, operation at this age carries such a high mortality rate that surgical *correction is seldom* warranted Certainly, if the *infant is* gaining weight and growing, neither cardiac enlargement nor pulmonary hypertension is an indication *for operation* Pulmonary hypertension at this age is due primarily to the excessively *large pulmonary* blood flow, it is mainly compensatory and still reversible

In children between three and six years of age, the existence of moderately severe pulmonary hypertension (that is, a systolic pressure of 60 to 70 mm of mercury) which is sustained over a period of years, is an indication for operation It is highly desirable to operate while the pulmonary pressure is appreciably lower than the systemic pressure and before irreversible changes develop in the pulmonary vascular bed

The crucial question, *and one which is not* easy to answer, is whether *or not* the pulmonary *pressure is going to fall* If there is clear demonstration of overriding of the *aorta*, *it is* almost certain that *the condition* will lead to progressive pulmonary vascular damage Under such circumstances operation in early childhood is clearly indicated If, on the other hand, there is evidence that the pulmonary pressure is falling, there is no urgency for operation

A pulmonary pressure of 40 mm of mercury is well tolerated for a long period of time Indeed, if over the years pulmonary hypertension is not progressive operation is not indicated for the sole purpose of the relief of that degree of pulmonary hypertension

In some instances even though the pulmonary pressure is low, *the excessive* pulmonary blood flow both starves the systemic circulation and *renders* the patient *extremely* susceptible to respiratory infections *For such* patients closure of the ventricular defect *is of great benefit*

Broadly speaking, the child who can be *most* benefited by operation is the

one with a large left to-right shunt, in whom the increased pressure in the pulmonary artery is due to the increased volume of blood in the pulmonary circulation and not to intimal changes in the lungs Such a child usually shows a harsh systolic murmur and a relatively normal pulmonic second sound, cardiac enlargement and increased vascular markings in the x ray, and electrocardiographic evidence of left ventricular dominance

It has been the repeated experience of all surgeons that the best results are obtained when there is a large left to-right shunt and when the pulmonary pressure, although elevated, is less than the systemic pressure, preferably under 70 per cent of systemic pressure, and the pulmonary vascular resistance is not fixed" *The effect of the inhalation of oxygen upon the volume of the shunt* and the pressure in the pulmonary artery is of great aid in the evaluation of the state of the pulmonary vascular bed If the volume of the shunt is increased or the pulmonary pressure drops, it is an indication of a flexible pulmonary vascular bed, hence there is reason to believe that the patient will be benefited by surgery

SPECIAL SURGICAL CONSIDERATIONS

The two most commonly associated conditions, which are easily overlooked and may cause difficulty at surgery, are patency of the ductus arteriosus and a persistent left superior vena cava which opens into the coronary sinus Anomalies of the aortic and mitral valves also require careful consideration

Patency of the ductus arteriosus may be difficult to diagnose in the presence of pulmonary hypertension Indeed, only if the catheter is passed through the ductus to the descending aorta can the condition be established with certainty Therefore it behooves the surgeon to explore the region of the ductus before placing the patient on a by pass, as in the presence of persistent patency of the ductus arteriosus a tremendous volume of blood may be shunted from the descending aorta to the lungs and returned to the left side of the heart

A left superior vena cava which opens into the coronary sinus may also cause trouble at operation If, in addition to the left superior vena cava, the innominate vein is present, the left superior vena cava can be ligated In the absence of an innominate vein it may be necessary to canulate the coronary sinus as well as the venae cavae

Aortic insufficiency may also cause difficulty because the aortic valve may be herniated into the septal defect Dr Henry Bahnson has shown that it is usually possible to repair the aortic valve after the ventricular septal defect has been

Abnormalities of the mitral and tricuspid valves call for special consideration Such anomalies may place an intolerable load upon the heart after closure of the ventricular septal defect *Acquired mitral stenosis* may be readily relieved by surgery but *congenital mitral stenosis* poses a more difficult problem On the other hand, *acquired mitral insufficiency* is difficult to correct, whereas *congenital mitral insufficiency*, when due to a cleft in the mitral valve, has been shown by Kirklin and others to be relatively easy to correct Furthermore, Kirklin[18] has found that the correction of a mitral insufficiency is quite as important when it occurs in combination with a ventricular septal defect as when it occurs with an ostium primum defect

Tricuspid insufficiency combined with a ventricular septal defect is rare but does occur Figure xxiv-7 illustrates a ventricular septal defect of the atrioventricular canal type in which the tricuspid valve was cleft but the mitral valve was normal Tricuspid insufficiency requires surgical correction quite as much as does mitral insufficiency

Multiple ventricular septal defects present a special problem Although the individual defects may be small and difficult to find, together they may place an intolerable burden on the heart Hence such defects must be searched for with great care and all of them must be closed

The development of complete heart block due to injury of the conduction system at operation is an ever present danger in the surgical closure of defects in the ventricular septum Complete atrioventricular dissociation acquired during surgery is far more serious than the development of complete heart block from acquired disease The latter may be compatible with life for many years, whereas the former is almost always fatal Indeed, even if the patient recovers from the operation and is apparently able to adjust to the slow heart rate, within a *period* of months, or at most a few years, sudden death may occur Consequently this danger must be considered in the evaluation of the risk versus the benefits of operation

Successful closure of a ventricular septal defect restores the heart and circulation to normal Although there is seldom an *immediate* reduction in the size of the heart during the ensuing year or two there is usually *a marked decrease in* its size (see Figures xxiv-11 and 13) If the defect is closed in childhood, *there is* every reason to hope that the pulmonary vascular bed will expand and the pulmonary pressure will return to normal

It is not yet known how smoothly the endocardium heals, therefore, even after uccessful operation prophylactic antimicrobial agents are still indicated prior to dental extraction or tonsillectomy Nevertheless, the restoration of the

circulation to normal, combined with the reduction in susceptibility to pulmonary infection, is a tremendous boon to the child

The resumption of activity should be more gradual for these patients than for those in whom the ventricle has not been opened Most of these patients are able to resume a relatively normal life within six months after operation The full benefit from the closure of a ventricular septal defect, however, is not reached until a year or a year and a half after operation

PROGNOSIS

Without surgery the prognosis depends mainly upon the height of the pulmonary pressure The high pressure with which the blood is pumped into the pulmonary artery leads to progressive injury to the pulmonary vascular bed The development of cyanosis is of bad prognostic import It indicates that changes in the lungs have become so extensive that there is a right to left shunt which becomes of steadily increasing magnitude Thereafter both anoxemia and polycythemia increase and the duration of life is limited

The later the onset of cyanosis and the more gradual the development of polycythemia, the better is the prognosis This is vividly illustrated by a patient whom the author first saw when he was forty years old At that time he gave the history that cyanosis had not been apparent until he was thirty five years of age His red blood cell count was six million per cu mm and his hematocrit reading was 61 per cent The oxygen saturation of his arterial blood was between 80 and 84 per cent Cardiac catheterization showed that the systolic pressure in his pulmonary artery was 100 mm of mercury, but the diastolic pressure was only 40 mm of mercury The author saw him again when he was fifty years old His condition was virtually unchanged Neither the red blood cell count nor the hematocrit reading had risen This sequence of events suggests that diastolic pressure in the pulmonary artery, which reflects the resistance in the lungs, is more important than the systolic pressure and that a high pulmonary systolic pressure may be well tolerated for many years provided the diastolic pressure remains low

Most patients, however, with an Eisenmenger complex die from hemoptysis or cardiac failure between the ages of twenty and forty years A few die from subacute bacterial endocarditis or from brain abscesses

Surgical closure of the ventricular defect restores the heart and the circulation to normal Thereafter the prognosis is excellent

SUMMARY

Large ventricular septal defects and hearts of the Eisenmenger type are characterized by excessive pulmonary blood flow and high pulmonary pressure During infancy the shunt is from left to right but, as the pulmonary resistance increases, a right to-left shunt may develop The malformation does not usually [illegible] early childhood Patients with this malforma[illegible]

[illegible]

[illegible] tured These patients suffer from excessive pulmonary blood [illegible] hypertension The progressive increase in the pulmonary hypertension increases the right to-left shunt and leads to the late development of cyanosis

The heart is usually but slightly enlarged, both ventricles share in the enlargement, and there may be enlargement of the left auricle, the right auricle is, however, of normal size Usually there is a harsh systolic murmur and a thrill along the left sternal border and the pulmonic second sound is accentuated and reduplicated

The x ray commonly shows fullness of the pulmonary conus and fluoroscopy shows a hilar dance

The electrocardiogram may show either a right or a left axis deviation and evidence of combined hypertrophy Evidence of left ventricular dominance is a favorable sign

Cardiac catheterization will show that the major shunt is from the left ventricle to the right ventricle The pressure in both the right ventricle and the pulmonary artery may be abnormally high it may approach but never exceeds the systemic pressure

When an Eisenmenger complex is suspected in young children, cardiac catheterization is indicated, as this is the time for surgery Although the development of cyanosis at or about the time of puberty is characteristic of this malformation, it is to be hoped that the diagnosis will be made before the patient becomes cyanotic

In infancy and early childhood a large ventricular septal defect requires differentiation from other malformations with a large left to-right shunt, such as persistent patency of the ductus arteriosus before the development of a continuous murmur, anomalies of the pulmonary venous return, an auricular septal defect an ostium primum defect or a persistent ostium atrioventriculare commune After the advent of cyanosis the malformation may resemble a pure pulmonary stenosis with persistent cyanosis, primary pulmonary hypertension, or occasionally a tetralogy of Fallot

Surgical closure of a ventricular septal defect is now possible with open heart surgery The principal indications for closure of a ventricular septal defect are cardiac enlargement, a predominant left to right shunt, and moderately severe pulmonary hypertension If the pulmonary pressure is equal or nearly equal to the systemic pressure, early operation (between three and six years of age) is indicated, because the high pressure with which blood is ejected to the lungs may cause irreparable injury to the pulmonary vascular bed Without surgical correction at an early age, such a malformation may lead to cyanosis, polycythemia, and hemoptysis, death usually occurs in early adult life from severe hemoptysis, brain abscess, or cardiac failure

If the pulmonary pressure is moderately elevated and the heart is enlarged, operation is relatively safe and virtually restores the heart and the circulation to normal

The prognosis for a large ventricular septal defect with moderate pulmonary hypertension is relatively good but is greatly improved by operation If the patient has severe pulmonary hypertension, the prognosis is guarded unless the condition is corrected by surgery

C *Defects between the Left Ventricle and the Right Auricle*

Defects in the ventricular septum which open into the right auricle are the rarest of all ventricular septal defects The anomaly was, however, mentioned by Meyer in 1857 He credits the first case to Buhl [14] Another case was reported by Hillier[15] in 1859 Nevertheless, when Perry et al [16] reported their case in 1949, they found only five cases in the literature Stahlman et al [17] have reported another case

NATURE OF THE MALFORMATION

The essential feature of the malformation is a defect in the left ventricle which opens into the right auricle Edwards[18] has shown that such defects may be of two types in the one a defect in the ventricular septum opens directly into the right auricle above the tricuspid valve, in the other a defect which lies in the lower portion of the membranous septum opens first into the right ventricle behind the septal leaflet of the tricuspid valve and then into the right auricle through a second defect in the tricuspid valve, which has become closely adherent to the septum Thus the shunt may be from the left ventricle into the right auricle, or there may be a shunt both at the ventricular level and at the auricular level The cross sectional diagram in Figure XXIV-18 shows the anatomical rela

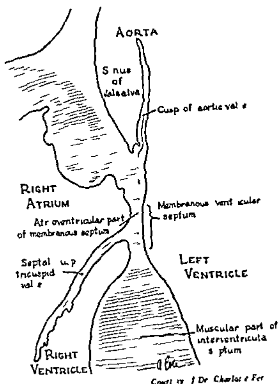


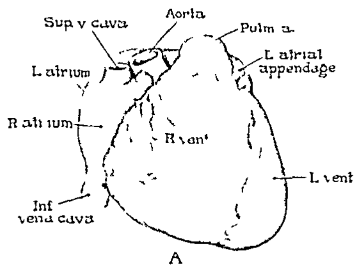
FIGURE XXIV–18 Cross section of heart showing the relation of the membranous septum to the right auricle the aorta and both ventricles

tion of the right auricle and the tricuspid valve to the left ventricle and the aortic valve and how readily it is possible for a defect in the membranous septum to open into the right auricle instead of the right ventricle A heart with such a defect[19] is shown in Figure XXIV–19

Inasmuch as the defect lies in the atrioventricular portion of the membranous septum the abnormality not infrequently involves the mitral and tricuspid valves and may affect the outflow tract of either the right or the left ventricle Consequently the malformation may be associated with a wide variety of anomalies

COURSE OF THE CIRCULATION

The blood in the right auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs, where it is oxygenated, and returned in the normal manner by the pulmonary veins to the left auricle Thence



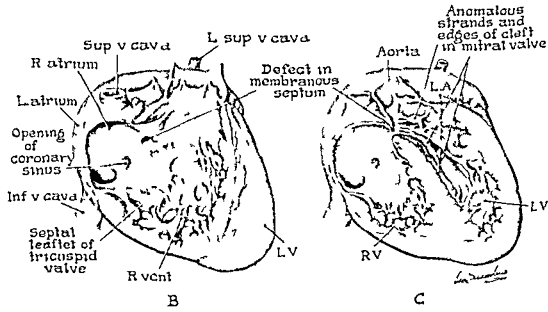


FIGURE XXIV-19 Defect in the membranous septum between the left ventricle and the right auricle

it flows to the left ventricle Most of the blood in the left ventricle is pumped out through the aorta to the body and returned in the normal manner by the superior vena cava and the inferior vena cava to the right auricle Inasmuch as there is a defect in the left ventricle which opens into the right auricle, with each ventricular systole some blood is pumped directly into the right auricle This oxygenated blood becomes mixed with the venous blood returned to the right auricle by the venae cavae Therefore it is a mixture of oxygenated and venous blood which flows into the right ventricle There the cycle starts again The course of the circulation is shown in Diagram xxiv–5

PHYSIOLOGY OF THE MALFORMATION

Inasmuch as the shunt is from the left ventricle to the right auricle, there is no question of dextroposition of the aorta Therefore it is the volume of the shunt which determines the pressure in the pulmonary artery Indeed, it affects the pressure in much the same manner as does an auricular defect The defect is smaller in size than is an auricular septal defect but the blood is shunted into the right auricle under systemic pressure Indeed, at surgery a jet of blood can be felt entering the right auricle Because of the force with which the blood is directed to the right auricle, the volume of the shunt is large

Any other alteration in the hemodynamics is due to additional associated malformations

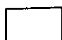
CLINICAL FINDINGS

The clinical findings vary with the exact location of the defect and the nature of the associated anomalies

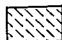
When the anomaly occurs as an isolated malformation, the clinical findings are in no way different from those which occur in other ventricular septal defects In other instances the clinical picture is that of the basic anomaly with which it is associated

Thus, in a case reported by Dammann [9] the clinical picture was that of a tetralogy of Fallot with infundibular stenosis and a large pulsating liver The pulsations at the margin of the liver gave the clue to the existence of some complicating anomaly and led to angiocardiography Angiocardiography showed evidence of a shunt from the right auricle to the left ventricle in auricular systole and a shunt from the left ventricle to the right auricle in ventricular systole Thus in retrospect, the information derived from the angiocardiographic studies was sufficient to establish the diagnosis

Diagram XXIV–5



Arterial blood (fully saturated)



Small admixture of venous blood
No visible cyanosis



Venous and arterial blood
Cyanosis visible



Venous blood

DIAGRAM XXIV-5

Septal defect between the left ventricle and the right auricle

In this malformation there is a ventricular septal defect which lies high up beneath the aortic valve and opens into the right auricle either directly or through a perforation in the tricuspid valve where this valve becomes plastered against the ventricular septum Although the opening is small blood is directed from an area of high pressure to an area of low pressure hence the volume of the shunt may be relatively large. The right auricle receives venous blood from the superior vena cava and the inferior vena cava and some oxygenated blood from the left ventricle This mixture of oxygenated and venous blood flows into the right ventricle and is pumped out through the pulmonary artery to the lungs where it is fully oxygenated The oxygenated blood is returned by the pulmonary veins to the left auricle thence it flows to the left ventricle A small amount of blood is pumped through the defect to the right auricle but most of the blood in the left ventricle is pumped out through the aorta to the systemic circulation and is returned by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again

Clinical diagnosis This defect involves the so-called critical area" of the membranous septum and the endocardial cushions Hence it is usually associated with some other serious malformation such as an aortic stenosis or a tetralogy of Fallot The signs and symptoms of the other defect may overshadow the signs of the ventricular septal defect When a patient has frank signs and symptoms of another malformation and catheterization reveals a step-up at the auricular level this malformation should be suspected A selective angiocardiogram with the dye placed in the left ventricle, should reveal the shunt to the right auricle

In the case reported by Ferencz[19] the primary difficulty was aortic stenosis, as the defect involved the upper portion of the ventricular septum, the aortic outflow tract, and the primordia of the atrioventricular valves

CARDIAC FINDINGS

The size and the shape of the heart depend upon the additional anomalies The heart is usually enlarged The enlargement involves the right auricle and the right ventricle

A systolic murmur and a thrill over the precordium are the rule

X RAY AND FLUOROSCOPIC FINDINGS

The heart is enlarged Usually both the right auricle and the right ventricle are enlarged and in addition the x ray shows fullness of the pulmonary conus and increased vascular markings

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram varies with the nature of the dominant malformation

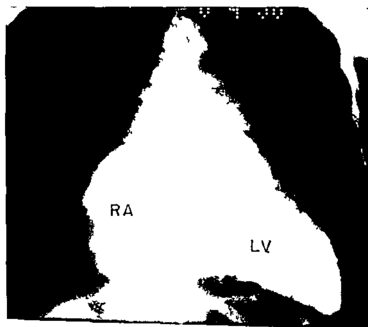
SPECIAL TESTS

Cardiac catheterization shows evidence of a left to-right shunt at the auricular level and there may or may not be an additional shunt at the ventricular level Although the defect is usually small, the difference between the pressure in the left ventricle and that in the right auricle is great, hence the volume of the shunt is relatively large

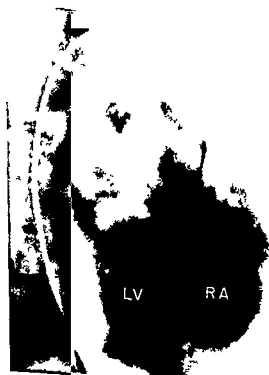
Angiocardiography is of aid in the diagnosis in that no evidence of a defect in the auricular septum is detected, but there may be evidence of a shunt from the right auricle to the left ventricle during auricular systole and from the left ventricle to the right auricle during ventricular systole, as in Dammann s case Morrow[21] has shown that by the injection of dye into the left ventricle it is possible to demonstrate the filling of the right auricle immediately after ventricular systole (see Figure XXIV–20)

DIAGNOSIS

The diagnosis is difficult, therefore simple awareness of the possibility is important The condition is to be suspected when the clinical findings are those of a ventricular septal defect and cardiac catheterization shows that the shunt is at the auricular level but angiocardiography reveals no evidence of such a de



Courtesy of Dr A G Morrow National Heart Institute



Courtesy of Dr A G Morrow National Heart Institute

FIGURE XXIV-20 Defect between the left ventricle and the right auricle

fect The diagnosis may be definitely established by a selective angiocardiogram taken after the dye has been injected into the left ventricle

DIFFERENTIAL DIAGNOSIS

An ostium primum type of auricular defect is the one most readily confused with this malformation The demonstration of a shunt at the auricular level, combined with failure to demonstrate an auricular septal defect, strongly suggests the malformation under discussion, provided that a partial anomaly of the pulmonary venous return has been excluded

A ventricular septal defect which involves the tricuspid valve (see Section B) could readily be confused with the malformation under discussion In the former malformation the shunt is mainly at the ventricular level, whereas in the malformation under discussion the shunt is at the auricular level and the pressure in the right ventricle does not approach that of the left ventricle

ASSOCIATED ANOMALIES

This defect is related to the formation of the endocardial cushions and the membranous septum Consequently the defect may occur with a wide variety of malformations which involve either the left side or the right side of the heart

TREATMENT

Although clinical diagnosis is difficult, surgical repair is relatively easy, as it does not require opening the ventricle A patch can be placed securely over the opening in the auricle and thereby the defect is closed

If a patient is operated upon for an auricular defect and the auricular septum is found to be intact, and yet a jet of blood can be felt coming through the auricular septum with each ventricular systole, it is almost certain that the defect lies between the left ventricle and the right auricle Once the defect is located, closure of the defect is relatively simple

PROGNOSIS

The prognosis varies with the size of the defect and the nature of the associated lesions If the defect occurs as an isolated malformation, it can be corrected by surgery

SUMMARY

A defect between the left ventricle and the right auricle is a rare type of ven

tricular septal defect, furthermore, it is frequently associated with other mal formations

The clinical findings are those of a ventricular septal defect Cardiac catheterization reveals a shunt at the auricular level and angiocardiography may reveal a communication between the right auricle and the left ventricle

The condition is of clinical importance because it can be corrected by surgery and thereby the heart and the circulation are restored to normal

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PROGNOSIS

The prognosis varies with the size of the defect and the nature of the associated lesions If the defect occurs as an isolated malformation, it can be corrected by surgery

SUMMARY

A defect between the left ventricle and the right auricle is a rare type of ven

CHAPTER XXV

ANEURYSM IN THE SINUS OF VALSALVA WITH RUPTURE INTO THE LESSER CIRCULATION

THE formation of an aneurysm in the right sinus of Valsalva which burrows through the wall of the heart and ruptures into the lesser circulation is generally considered to result from a congenital weakness in the wall at the base of the aorta For this reason, although both the formation and the rupture of the aneurysm occur after birth, the condition is classified as a congenital anomaly

Mall[1] in 1912 pointed out that aneurysms occur at points of weakness in the formation of the cardiac wall at the base of the aorta Abbott[2] in 1919 reported a case in which such an aneurysm had ruptured into the right ventricle Upon a review of the clinical history she realized that the signs and symptoms were so distinctive as to constitute a clinical syndrome To the best of the author s knowledge, Tompkins[3] was the first to make a correct clinical diagnosis In his case, as is so common when the aneurysms are due to acquired disease, the aneurysm was syphilitic in origin In a number of the earlier reports the aneurysm was caused by a mycotic infection in others, no evidence of infection was found Even when the aneurysm is the result of infection, its formation in this particular location is due to the existence of a congenital weakness at the site

With the advent of chemotherapy and antibiotics it is reasonable to expect that aneurysms due to infection will markedly diminish therefore over a period of years the group in which the abnormality is entirely the result of a congenital weakness in the wall of the aorta will become proportionally more common

A review of the development of the aortic septum as it meets the ventricular septum will help to clarify the nature of this malformation

EMBRYOLOGY

Mall[1] in his analysis of the formation of the aortic septum pointed out that, when the aortic septum divides the truncus arteriosus into the pulmonary artery and the aorta, the aorta shifts from the right side of the heart to occupy its normal posterior position opening into the left ventricle Simultaneously with the shift of the aorta from right to left, the inferior septum at the base of the ventricle which is destined to form the upper part of the ventricular septum, shifts

13 Kirklin, J W Personal communication

14 Quoted by H Meyer in Ueber angeborene Enge oder Verschluss der Lungenarterienbahn Virchows Arch f path Anat 12 497, 1857

15 Hillier, T Congenital malformation of the heart, perforation of the septum ventriculorum, establishing a communication between the left ventricle and the right auricle. Tr Path Soc London 10 110–111, 1859

16 Perry, E L, H B Burchell, and J E Edwards Cardiac clinics, congenital communication between the left ventricle and the right atrium, co-existing ventricular septal defect and double tricuspid orifice Proc Staff Meet, Mayo Clin 24 198–206, 1949

17 Stahlman, M, S Kaplan, J A Helmsworth, L C Clark, and H W Scott, Jr Syndrome of left ventricular–right atrial shunt resulting from high interventricular septal defect associated with defective septal leaflet of the tricuspid valve Circulation 12 813–818, 1955

18 Edwards, J E Congenital malformations of the heart and the great vessels *In* S E Gould (ed), Pathology of the Heart Springfield, Ill, Thomas, 1953

19 Ferencz, C Atrio-ventricular defect of membranous septum, left ventricular–right atrial communication with malformed mitral valve simulating aortic stenosis, report of a case Bull Johns Hopkins Hosp 100 209–222, 1957

20 Dammann, J F, Jr Personal communication

21 Morrow, A G Personal communication

is a high ventricular septal defect A drawing of such a case is shown in Figure xxv-1 Figure xxv-2 shows two photographs of a heart in which an aneurysm originating in the right sinus of Valsalva ruptured into the right auricle and a second sinus tract ruptured into the right ventricle

COURSE OF THE CIRCULATION

The course of the circulation prior to the rupture of the aneurysm is altered only if there is an additional malformation of the heart, when the heart is otherwise normal, the circulation of the blood *is normal* The rupture of the aneurysm establishes a communication between the aorta and the chamber into which the aneurysm ruptures This may be into either the right ventricle or the right auricle, occasionally the aneurysm ruptures into the pulmonary artery

The blood from the right auricle flows into the right ventricle and is pumped out by way of the pulmonary artery to the lungs The blood from the lungs is returned in the normal fashion to the left auricle, thence it flows to the left ventricle and *is pumped out through the aorta to the systemic circulation* With the closure of the semilunar valves, the high pressure at the base of the aorta in early diastole directs the blood into the coronary arteries and in this malformation into the aneurysmal sac which opens into the right side of the heart If the aneurysm has burrowed through the ventricular septum, the contraction of the ventricles during systole must markedly decrease if not completely close, the communication between the two sides of the heart The main flow of blood from the aorta to the lesser circulation occurs during diastole The course of the circulation when the aneurysm opens into the right ventricle is shown in Diagram xxv-1 If the aneurysm opens into the right auricle, some oxygenated blood from the aorta will enter the right auricle and thence flow into the right ventricle, as shown in Diagram xxv-2

PHYSIOLOGY OF THE MALFORMATION

The condition is essentially that of an arteriovenous aneurysm Inasmuch as the aneurysm is produced by the incessant pounding of the blood against a weakened arterial wall, the aneurysm tends to increase in size and thus places an ever increasing burden on the heart

CLINICAL FINDINGS

An abrupt onset of signs and symptoms is characteristic of the clinical syndrome These develop at the time of the rupture of the aneurysm The signs and

from left to right Eventually the right margin of the aortic septum fuses with the inferior septum to form the membranous septum If the aorta does not shift sufficiently, the aorta over rides the right ventricle as in the Eisenmenger complex If the aorta reaches its normal position, but the fusion between the aortic septum and the ventricular septum is not complete, there results a high ventricular septal defect Occasionally the development may progress until the aortic septum has been completely formed but a weakness in the arterial wall persists at the base of the right sinus of Valsalva Edwards and Burchell[4] have shown that the aortic wall immediately above this point may also be abnormally thin It is in this region that the wall gives way under the incessant impact of the heart beat, thereby an aneurysmal pouch is formed which burrows through the cardiac septum and eventually ruptures into the right side of the heart

NATURE OF THE MALFORMATION

The essential feature of this malformation is an aneurysm which originates at the base of the right sinus of Valsalva and burrows through the wall of the heart The aneurysm usually ruptures into the right ventricle, the exact point of rupture is subject to considerable variation Some aneurysms rupture high up on the septal wall just beneath the pulmonary orifice and some near the insertion of the tricuspid valve The latter are likely to cause distortion of the tricuspid valve and tricuspid insufficiency A number of cases have been reported[5-8] in which the aneurysm pushed its way horizontally and ruptured into the right auricle The author has seen one case in which the sinus tract divided and one branch ruptured into the right auricle and the other into the right ventricle, and a second case in which the rupture was into the right auricle Cases have been reported[9, 10] in which the aneurysm pushed upward and ruptured into the pulmonary artery It has been repeatedly emphasized[11-13] that all the aneurysms have one important feature in common namely, they are *intracardiac* aneurysms For this reason, although the rupture of the aneurysm causes an arteriovenous communication, the condition is not immediately fatal In contrast to such aneurysms, those which form in the aorta behind the aortic cusp, bulge exteriorly, and rupture into the pericardium are immediately fatal The same is true of those which rupture through the pericardium into the left pleural cavity It is only the intracardiac aneurysms which are not immediately fatal, that produce a distinctive clinical syndrome

When the aneurysm is due to a congenital weakness, the condition is frequently associated with other congenital anomalies, the most common of these



Viewed from left side of heart

[illegible]



Viewed from right side of heart

[illegible]

FIGURE XXI-2 Aneurysm in the sinus of Valsalva with rupture into the right auricle. Adult

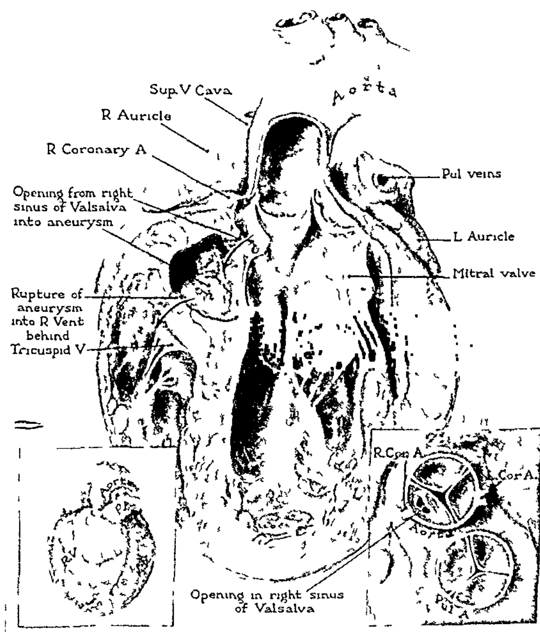


FIGURE XXV-1 Aneurysm in the sinus of Valsalva with rupture into the right ventricle at fourteen years of age

Diagram XXV–1

Aneurysm in the sinus of Valsalva with rupture into the right ventricle

This malformation consists of an aneurysm which originates in the right sinus of Valsalva, burrows through t[illegible] the ventricular septum, and eventually ruptures into the right ventricle T[illegible] lar septal defect. The con[illegible] of the aneurysm.

The blood from the right auricle passes into the right ventricle and is pumped out through the pulmonary artery to the lungs The oxygenated blood from the lungs is returned by the pulmonary veins to the left auricle Thence the blood passes into the left ventricle and is pumped out by way of the aorta At the end of systole, after closure of the aortic valves that portion of the blood from the aorta which enters the right sinus of Valsalva passes through the ruptured opening into the right ventricle. Thus oxygenated blood from the aorta mixes with the venous blood from the right auricle. This mixture of blood is again pumped around the pulmonary circulation Thus an arteriovenous shunt is established The shunt is from left to right, there is no clubbing or cyanosis

Inasmuch as the opening is aneurysmal in nature and the pressure in the systemic circulation remains higher than that in the pulmonary circulation, there is an increasing pressure upon the aneurysmal opening which inevitably causes progressive enlargement of the opening and an increasing burden on the heart The malformation usually leads to rapid progressive cardiac enlargement and death within a relatively short time after rupture

Clinical diagnosis The outstanding clinical feature is the abrupt onset of signs and symptoms which develop at the time of the rupture of the aneurysm There may be a history of sudden acute cardiac pain which is followed by signs of myocardial insufficiency On physical examination there is a continuous murmur over the precordium in the third and fourth left interspaces The murmur is similar to that of a patent ductus arteriosus but usually has a diastolic accentuation and seems to originate close to the chest wall It is maximal at a lower level than is usual with persistent patency of the ductus arteriosus After rupture there are signs of progressive cardiac failure and the duration of life is relatively brief

DIAGRAM XXV–1

Head and upper extremities
R Lung
L Lung
S V C
P A
Pul A
Aorta
P V
Pul V
Pulmonary Art
L A
R A
L V
R V
Descending Aorta
I V C
Sinus of Valsalva and Aneurysm Ruptured into Right Ventricle
Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis

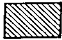

Venous blood

Diagram XXV-2

Aneurysm in the sinus of Valsalva with rupture into the right auricle

In this instance the aneurysm which originates in the right sinus of Valsalva burrows through the wall of the auricular septum and eventually ruptures into the right auricle There may or may not be a ventricular septal defect. The condition does not become clinically manifest until the rupture of the aneurysm

The blood from the right auricle passes into the right ventricle and is pumped out through the pulmonary artery to the lungs The oxygenated blood from the lungs is returned by the pulmonary veins to the left auricle. Thence the blood passes into the left ventricle and is pumped out by way of the aorta At the end of each systole, after closure of the aortic valves the blood from the aorta which enters the right sinus of Valsalva passes through the ruptured opening into the right auricle. There the oxygenated blood from the aorta meets the venous blood returned by the superior vena cava and the inferior vena cava to the right auricle This blood, with a relatively high oxygen content, passes into the right ventricle So the cycle continues Thus an arterio-venous aneurysm is established The shunt is always from left to right, there is no cyanosis

Inasmuch as the opening is aneurysmal in nature and the pressure in the systemic circulation is higher than that in the pulmonary circulation there is an increasing pressure upon the aneurysmal opening which usually causes progressive enlargement

[illegible]

a relatively short time after the rupture of the aneurysm

Clinical diagnosis The outstanding clinical feature is the abrupt onset of signs and symptoms which develop at the time of the rupture of the aneurysm Frequently there is a history of sudden acute cardiac pain which is followed by signs of myocardial insufficiency There is great enlargement of the heart to the right and to the left The murmur is similar to that of a patent ductus arteriosus but usually has a diastolic accentuation and seems to originate close to the chest wall It is maximal over the precordium and to the right of the sternum and not in the second left interspace. After the rupture cardiac failure develops and becomes severe Thereafter the duration of life is usually relatively brief

DIAGRAM XXV–2

Head and upper extremities

R Lung

L Lung

S.V.C.

P.A.

Aorta

Pul. A

PV

Pul V

Pulmonary Artery

L A

R.A.

Descending Aorta

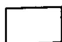
Aneurysm from Sinus of Valsalva ruptured into Right Auricle

L V

R V

I.V.C.

Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

DIAGRAM XXV–2

Aneurysm in the sinus of Valsalva with rupture into the right auricle

In this instance the aneurysm which originates in the right sinus of Valsalva burrows through the wall of the auricular septum and eventually ruptures into the right auricle. There may or may not be a ventricular septal defect. The condition does not become clinically manifest until the rupture of the aneurysm

The blood from the right auricle passes into the right ventricle and is pumped out through the pulmonary artery to the lungs The oxygenated blood from the lungs is returned by the pulmonary veins to the left auricle Thence the blood passes into the left ventricle and is pumped out by way of the aorta At the end of each systole after closure of the aortic valves, the blood from the aorta which enters the right sinus of Valsalva passes through the ruptured opening into the right auricle. There the oxygenated blood from the aorta meets the venous blood returned by the superior vena cava and the inferior vena cava to the right auricle This blood with a relatively high oxygen content, passes into the right ventricle So the cycle continues Thus an arterio-venous aneurysm is established The shunt is always from left to right, there is no cyanosis

Inasmuch as the opening is aneurysmal in nature and the pressure in the systemic

tion generally leads to progressive cardiac enlargement Death usually occurs within a relatively short time after the rupture of the aneurysm

Clinical diagnosis The outstanding clinical feature is the abrupt onset of signs and symptoms which develop at the time of the rupture of the aneurysm Frequently there is a history of sudden acute cardiac pain which is followed by signs of myocardial insufficiency There is great enlargement of the heart to the right and to the left. The murmur is similar to that of a patent ductus arteriosus but usually has a diastolic accentuation and seems to originate close to the chest wall It is maximal over the precordium and to the right of the sternum and not in the second left interspace. After the rupture cardiac failure develops and becomes severe Thereafter the duration of life is usually relatively brief

symptoms vary according to the size and the location of the aneurysm, the chamber into which the aneurysm ruptures, and whether or not the condition is associated with another malformation of the heart

The age at which rupture may occur varies with the etiology Aneurysms which are congenital in origin usually rupture in early adult life, between the ages of fourteen and thirty,[2] whereas those which are due to syphilis commonly occur at a slightly later age Inasmuch as the abnormality does not become clinically manifest until the rupture of the aneurysm, this clinical syndrome never occurs in infants and young children

Pain may or may not be an outstanding feature at the time of rupture The history is usually that of a person who has previously been in excellent health and suddenly develops signs and symptoms of myocardial insufficiency Although some patients give a history of excruciating precordial pain, it is striking that such a history is not always obtainable The author has seen four patients with this condition in whom no history of pain could be elicited Schuster[14] and Chipps[15] have observed that severe pain combined with the symptoms of coronary occlusion may occur if the aneurysm involves the coronary orifice

Dyspnea generally develops abruptly after the rupture of the aneurysm and becomes progressively more severe

Cyanosis is absent The flow of blood is from the aorta to the lesser circulation and the structure of the aneurysmal opening precludes the possibility of a reversal in the direction of the shunt Therefore cyanosis can result only from peripheral stasis or from an associated malformation

The pulse pressure is wide The diastolic pressure is low and there may be peripheral signs of aortic insufficiency due to the reflux of blood from the aorta into the right side of the heart during diastole

The liver frequently becomes severely congested Rapid enlargement of the liver is common when the aneurysm ruptures into the right ventricle in such a manner as to involve the tricuspid valve Under such circumstances pulsations at the margin of the liver are often readily palpable

Congestion in the lungs occurs with cardiac failure but is less severe than is the engorgement of the liver

Edema and anasarca may be outstanding terminal difficulties

CARDIAC FINDINGS

The heart is usually enlarged The rupture of the aneurysm causes an abrupt alteration in the circulation in which a left to-right shunt is established This

sudden change in the hemodynamics causes an abrupt increase in the size of the heart, which may be so great as to precipitate cardiac failure Although the patient may adjust to this sudden increase in the load placed on the heart and regain compensation, the aneurysm tends to increase in size Sooner or later the heart usually undergoes progressive enlargement which eventually leads to in tractable cardiac failure

A continuous murmur and a thrill throughout systole and diastole are the most characteristic of all the findings In most instances the murmur has the quality of the continuous murmur produced by patency of the ductus arteriosus but is of maximal intensity in the third or fourth interspaces close to the sternum It is usually louder to the left than to the right of the sternum but varies with the point of rupture of the aneurysm It has been repeatedly emphasized that the murmur appears to originate from a superficial portion of the heart In most instances, the murmur sounds close to the ear It may be so loud that it can be heard by the unaided ear two inches from the chest The murmur is widely transmitted over the precordium and may be audible over the entire chest, it is maximal over the heart, not in the second left interspace beneath the clavicle Thus the location of the murmur differs from that of a patent ductus arteriosus

The diastolic component of the murmur is due to the flow of blood during diastole from the aorta to the area of low pressure on the right side of the heart Abbott[4] and Gressner[16] have commented on its curious swishing quality The diastolic murmur more closely resembles that produced by patency of the ductus arteriosus than by that of aortic insufficiency The murmur differs from the con tinuous murmur produced by persistency of the ductus arteriosus in that the diastolic component is usually louder than the systolic component, whereas in a patent ductus arteriosus the systolic component is generally the louder of the two

The origin of the continuous murmur is obvious when the aneurysm ruptures into the right auricle, because during both systole and diastole blood is forced from the aorta into an area of low pressure The volume of blood so shunted may be enormous When the aneurysm ruptures into the right ventricle, the contraction of the ventricle must lessen or prevent the flow of blood during systole In such instances, it seems probable that the systolic murmur is caused by a ventricular septal defect, which is relatively common in association with this anomaly The author has followed two patients who over a period of years had evidence of a ventricular septal defect, both patients abruptly developed a continuous murmur and signs of rupture of an aneurysm of the sinus of

Valsalva into the lesser circulation Indeed, in one of these patients, the alteration in the hemodynamics was so marked that the mother noted the increased activity over the boy's precordium and upon palpation realized that the murmur and thrill had changed, and for that reason she again brought her son to the doctor On examination the patient was found to have developed a superficial continuous murmur over the lower precordium In this patient, as well as in the other, the systolic murmur clearly antedated the diastolic murmur and must have had a different origin Nevertheless, regardless of origin, the occurrence of a murmur in both systole and diastole is the rule

Tricuspid insufficiency is a common complication when the aneurysm ruptures into the right ventricle, because the usual point of rupture is close to the septal leaflet of the tricuspid valve The leaflet becomes inflamed and distorted and the valve becomes insufficient In such instances the tricuspid insufficiency may play a role in the production of the systolic murmur The feature of diagnostic significance in tricuspid insufficiency, however, is not the systolic murmur but the engorgement of the liver, and above all the pulsations palpable at the margin of the liver

Rapid progression of signs and symptoms is the rule In spite of all therapy the heart continues to enlarge and the patient develops increasingly severe decompensation Death results from cardiac failure The condition is frequently fatal within two or three months after the rupture of the aneurysm

It is, however, noteworthy that the patient may regain compensation and live for a number of years Abbott[2] reported one patient who lived for nine years after rupture of the aneurysm The author has seen one patient in whom the clinical diagnosis became clearly apparent immediately after she developed cardiac failure in January, 1952, and who was still living in January, 1960 These observations indicate that the condition is not always as rapidly fatal as has hitherto been believed

X RAY AND FLUOROSCOPIC FINDINGS

The x ray findings are subject to wide variation, depending upon the associated malformation and whether the aneurysm ruptures into the right auricle, the right ventricle, or the pulmonary artery Albrecht[17] has reported instances in which the aneurysm projected toward the base of the aorta and was visible in the x ray Most aneurysms, as previously mentioned, lie within the septal wall and hence rarely cause any distortion of the cardiac silhouette Enlargement of the right auricle is usually greater and occurs earlier when the aneurysm rup

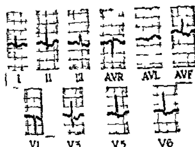


FIGURE XXV-3 Aneurysm in the sinus of Valsalva with rupture into the right auricle

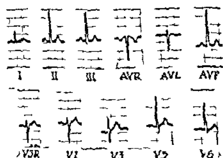


FIGURE XXV-4 Aneurysm in the sinus of Valsalva with rupture into the right ventricle

tures into the right auricle than when it ruptures into the right ventricle Only in the rare instances when the aneurysm ruptures into the pulmonary artery are pulsations in the hilar vessels likely to be seen

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram may show an intraventricular block Snyder and Hunter[18] believe that the block is caused by the extension of the aneurysm through the ventricular septum and its encroachment upon the main branches of the bundle of His Micks[19] has reported a case in which successive electrocardiograms taken over a short period of time showed that the intraventricular block was progressive In many instances, however, no evidence of conduction disturbance is found (see Figures XXV-3 and 4) Thus the electrocardiogram may or may not be of diagnostic help

SPECIAL TESTS

The red blood cell count the amount of available hemoglobin and *the hematocrit reading* are within normal limits

The circulation time is usually prolonged because of cardiac failure

Cardiac catheterization, except in the rare instances in which the aneurysm ruptures into the pulmonary artery, is of diagnostic aid Although the clinical findings may suggest patency of the ductus arteriosus or possibly aortic insufficiency, cardiac catheterization shows that the increase in the oxygen content of the blood does not occur in the pulmonary artery but in the right ventricle or in the right auricle, depending upon the point of rupture

Angiocardiography may offer confirmatory evidence as to the nature of the malformation When the aneurysm opens into the right auricle there may be an initial dilution of dye in that chamber and in addition the right auricle may be revisualized at the time the aorta fills

Aortography theoretically should be of diagnostic aid by the demonstration of the immediate reflux of dye from the base of the aorta into the right auricle or the right ventricle Such a phenomenon could occur only if there were a communication between the aorta and the right side of the heart In the only instance in the author s experience in which aortography was attempted, the catheter, inserted into the aorta for the injection of the dye, slipped from the base of the aorta into an area of low pressure The doctors were so astonished and alarmed that no samples were taken, the catheter was immediately withdrawn and the procedure terminated Nevertheless, the entrance of the catheter into an area of low pressure almost certainly indicated that it had been carried from the aorta into the right auricle by the regurgitant stream of blood The diagnosis is not proven, as the patient is still living She had, however, abruptly developed signs of cardiac failure and was found to have a loud continuous murmur best heard to the right of the sternum in the third and fourth interspaces Furthermore, upon cardiac catheterization the increase in the oxygen content of the blood was found to occur in the right auricle These findings taken together are strongly suggestive of an aneurysm in the right sinus of Valsalva which has ruptured into the right auricle

DIAGNOSIS

The diagnosis is based upon the findings of a loud continuous murmur with a diastolic accentuation which has the quality but not the location of the murmur produced by patency of the ductus arteriosus in a patient who is suffering from dyspnea or severe cardiac failure It is the combination of the cardiac enlargement, the sudden onset of signs and symptoms, the character and the location of the murmur which gives the clue to the diagnosis The diagnosis may be confirmed by cardiac catheterization in that the increase in the oxygen con

ent of the blood occurs in the auricle or the ventricle, and not in the pulmonary artery It may also be confirmed by aortography

DIFI

[illegible] continuous murmur over [illegible] tions in which there is a continuous murmur over [illegible] from other causes of progressive cardiac failure

Persistent patency of the ductus arteriosus is differentiated primarily by the location of the murmur The murmur of a patent ductus arteriosus is maximal above the heart in the second left interspace and the murmur and the thrill are not strikingly superficial The murmur of a patent ductus arteriosus usually has a systolic accentuation In the malformation under discussion the diastolic element is the louder of the two components In addition pulsations in the hilar shadows never occur except possibly when the aneurysm ruptures into the pulmonary artery

Syphilitic heart disease with aortic insufficiency may be confused with a ruptured aneurysm of the sinus of Valsalva as the occurrence of a murmur in diastole associated with a wide pulse pressure suggests the possibility of an aortic insufficiency In a ruptured aneurysm however the murmur is much more superficial and much more rasping than is usual with an aortic insufficiency Furthermore cardiac catheterization shows evidence of a left to-right shunt

Aneurysms of the aorta and *coronary thrombosis* may cause pain and cardiac insufficiency but such conditions are not usually associated with murmurs and thrills

Pericarditis may occasionally be confused with the malformation under discussion because of the occurrence of a superficial murmur in both systole and diastole The murmur associated with a ruptured aneurysm is, however, far harsher than that of a pericarditis and is always of maximal intensity in the third interspace to the right or to the left of the sternum whereas a pericardial friction rub is seldom both extremely loud and sharply localized

Anomalous communication between the right coronary artery and the right auricle or the right ventricle may give a similar clinical picture Indeed, this rare anomaly as shown by Espino-Vela et al,[9] may give the same clinical picture as a rupture of the sinus of Valsalva except that the condition dates from birth or is first detected at an early age Furthermore there is no sudden change of signs and symptoms and the condition is not progressive The murmur is usually detected upon routine examination It simulates that of a patent ductus

Cardiac catheterization except in the rare instances in which the aneurysm ruptures into the pulmonary artery, is of diagnostic aid Although the clinical findings may suggest patency of the ductus arteriosus or possibly aortic insufficiency, cardiac catheterization shows that the increase in the oxygen content of the blood does not occur in the pulmonary artery but in the right ventricle or in the right auricle, depending upon the point of rupture

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DIFFERENTIAL DIAGNOSIS

The malformation under discussion is to be differentiated from other conditions in which there is a continuous murmur over the body of the heart and from other causes of progressive cardiac failure

Persistent patency of the ductus arteriosus is differentiated primarily by the location of the murmur The murmur of a patent ductus arteriosus is maximal above the heart in the second left interspace and the murmur and the thrill are not strikingly superficial The murmur of a patent ductus arteriosus usually has a systolic accentuation In the malformation under discussion the diastolic element is the louder of the two components in addition pulsations in the hilar shadows never occur except possibly when the aneurysm ruptures into the pulmonary artery

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Aortography theoretically should be of diagnostic aid by the demonstration of the immediate reflux of dye from the base of the aorta into the right auricle or the right ventricle Such a phenomenon could occur only if there were a communication between the aorta and the right side of the heart In the only instance in the author s experience in which aortography was attempted, the catheter, inserted into the aorta for the injection of the dye, slipped from the base of the aorta into an area of low pressure The doctors were so astonished and alarmed that no samples were taken, the catheter was immediately withdrawn and the procedure terminated Nevertheless, the entrance of the catheter into an area of low pressure almost certainly indicated that it had been carried from the aorta into the right auricle by the regurgitant stream of blood The diagnosis is not proven, as the patient is still living She had, however, abruptly developed signs of cardiac failure and was found to have a loud continuous murmur best heard to the right of the sternum in the third and fourth interspaces Furthermore, upon cardiac catheterization the increase in the oxygen content of the blood was found to occur in the right auricle These findings taken together are strongly suggestive of an aneurysm in the right sinus of Valsalva which has ruptured into the right auricle

DIAGNOSIS

The diagnosis is based upon the findings of a loud continuous murmur with a diastolic accentuation which has the quality but not the location of the murmur produced by patency of the ductus arteriosus in a patient who is suffering from dyspnea or severe cardiac failure It is the combination of the cardiac enlargement, the sudden onset of signs and symptoms, the character and the location of the murmur which gives the clue to the diagnosis The diagnosis may be confirmed by cardiac catheterization in that the increase in the oxygen con

TREATMENT

Digitalis and supportive therapy may give temporary relief but no amount of medical treatment can heal a ruptured intracardiac aneurysm Therefore medical treatment will seldom fundamentally alter the course of events

Surgical closure of the aneurysmal opening at the base of the aorta may cure the condition The author knows of one woman in whom the rupture of an *aneurysm in the sinus* of Valsalva occurred during the stress of delivery Thereafter she developed severe cardiac failure with edema and ascites Dr Henry Bahnson [1] operated on this patient and was able to close the orifice of the aneurysm, which had ruptured into the right auricle, and also to re-enforce the base of the aorta Since then the woman has enjoyed good health Nevertheless, even after successful surgery the patient should be warned against strenuous or sudden exertion, as the base of the aortic wall is abnormally weak and could again give way

PROGNOSIS

The prognosis is extremely serious Usually there is a progressive extension of the aneurysm The size and the location of the aneurysm may, however, be such as to be compatible with life for a number of years Abbott reported a case in which there was strong evidence that the patient lived for nine years after the rupture of the aneurysm This, however is the exception Progressive circulatory failure is a conspicuous feature of the clinical syndrome Without surgery the patient usually dies within a few months after the onset of cardiac symptoms, that is shortly after the rupture of the aneurysm Therefore, except in the rare instances in which the patient responds to medical treatment, surgical treatment is strongly indicated Successful closure of the aneurysm is life saving and there is reason to hope that there will be no recurrence of the condition

SUMMARY

An aneurysm which originates in the right sinus of Valsalva and ruptures into the lesser circulation is the result of a congenital weakness in the aortic wall at the base of the heart The aneurysm may be caused by syphilis or by a mycotic infection or may result merely from the incessant pounding of blood against a point of weakness in the cardiac septum

The condition is characterized by the sudden onset of signs and symptoms

arteriosus, except that its location is maximal in the third or fourth left interspaces and not at the base of the heart Cardiac catheterization will reveal the increase in the oxygen content of the blood to occur in the auricle or the ventricle, as the case may be, and not in the pulmonary artery The age of the patient and the absence of symptoms give the clue to the diagnosis (see Chapter xxix)

Congenital arteriovenous aneurysm of the coronary artery produces a superficial murmur over the precordium The clinical history is entirely different in that there is no sudden onset of signs and symptoms On the contrary, the condition is usually asymptomatic for a number of years Angiocardiography clearly reveals the multiple aneurysms over the ventricle (see Chapter xxix)

Congenital aneurysmal dilatation of the coronary artery may cause a superficial continuous murmur over the body of the heart (see Chapter xxix) This vessel may open into the right ventricle Aortography shows the greatly dilated, tortuous coronary artery (see Figure xxix–12)

Primary pulmonary hypertension combined with pulmonary insufficiency may occasionally be confused with the malformation under discussion because in the presence of cardiac failure the murmur may simulate the superficial murmur of an aneurysm which has ruptured from the sinus of Valsalva into the right side of the heart Treatment with digitalis differentiates the two conditions, because on restoration of compensation the continuous murmur is replaced by the murmur characteristic of pulmonary insufficiency

Hemi truncus arteriosus in an otherwise normal heart gives a continuous murmur with an abnormal location Usually the murmur is maximal over the right or the left lung and not over the precordium (see Chapter xii, Section B), it is more likely to be confused with patency of the ductus arteriosus than with a rupture of an aneurysm of the sinus of Valsalva

Localized constriction of the pulmonary artery may cause a continuous murmur Constriction in the pulmonary artery, however, causes a continuous murmur over the lungs, not over the precordium and the cardiac findings, if present, are those of right sided hypertrophy

COMMONLY ASSOCIATED MALFORMATION

A small ventricular septal defect is of frequent occurrence in patients in whom an aneurysm develops in the right sinus of Valsalva Such a ventricular septal defect is usually a small one which lies in the membranous septum and in itself causes no difficulty

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21 Bahnson H T Personal communication

in a young individual who has previously enjoyed good health Precordial pain and dyspnea appear early Cyanosis is absent

Examination of the heart reveals a harsh continuous murmur which has the quality of the murmur of a patent ductus arteriosus but is maximal over the body of the heart, the murmur appears to originate just beneath the chest wall and usually has a diastolic accentuation The thrill feels very superficial, as if it had originated close to the hand

The condition generally leads to rapid, progressive cardiac failure with dyspnea and edema and often with evidence of tricuspid insufficiency and anasarca

The diagnosis is based upon the occurrence of a superficial continuous murmur over the precordium in the third and fourth interspaces close to the sternum

The diagnosis is substantiated by cardiac catheterization which reveals a left to-right shunt proximal to the pulmonary artery or by aortography which reveals dye in the right auricle or the right ventricle immediately after the aorta is filled

The condition requires differentiation from persistent patency of the ductus arteriosus, from aortic insufficiency, and occasionally from primary pulmonary hypertension combined with pulmonary insufficiency It also may require differentiation from an aneurysm of the aorta, coronary occlusion, and pericarditis The condition may be simulated by an anomalous communication between the right coronary artery and the right ventricle and also by a coronary arteriovenous aneurysm, by an aneurysmal dilatation of the coronary artery, and possibly by a hemi truncus arteriosus or by a localized constriction of the pulmonary artery

Medical treatment may restore and maintain compensation for a period of weeks or months and occasionally for years It however does not alter the underlying abnormality Therefore without surgery prognosis is extremely serious Surgical closure has been successfully accomplished Such treatment cures the condition and thereby completely alters the prognosis

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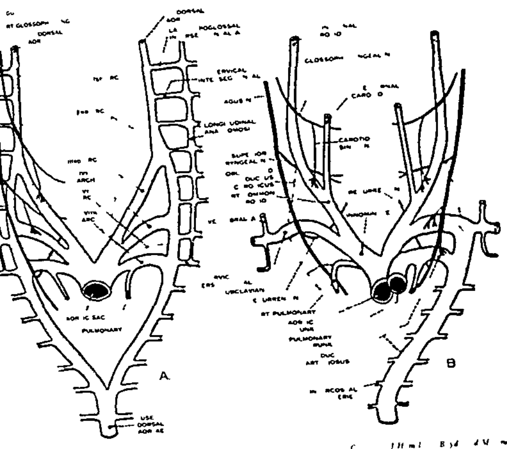


FIGURE XXVI 1 (A) Branchial arch arteries and (B) origin of the aorta and the pulmonary artery

is concerned with these anomalies Section c discusses other anomalies of systemic vessels as they arise from the aortic arch

A Right Aortic Arch

EMBRYOLOGY

The aorta is developed from three primitive structures the aortic sac, the fourth branchial arch and two dorsal aortae which fuse together and lie to the left of the vertebral column The pulmonary artery and the ductus arteriosus are derived from the sixth branchial arch Under normal conditions it is the fourth *left* branchial arch which forms the arch of the aorta and the fourth arch on the right side which atrophies and disappears (see Figure XXVI-1B) In the anomaly

CHAPTER XXVI

ANOMALIES OF THE AORTIC ARCH

ANOMALIES in the direction in which the aorta arches or abnormalities in the origin of the great vessels from the arch of the aorta are by no means rare; they may occur together or separately.

EMBRYOLOGY

Congdon[1] in his extensive studies of the embryology of the aortic arch has shown how delicate and complex is the development of the great vessels and the major vascular pattern. Figure XXVI–1 shows the primitive vascular bed and the origin of the aorta and the pulmonary artery.[2] According to Streeter,[3] the blood vessels develop in response to the stress and strain placed on the primitive vascular bed. If this is true, it is easy to understand how a slight variation in the rotation or position of the heart or in the formation of the ventricles may lead to anomalies in the development of the great vessels. Figure XXVI–2 is a schematic representation of the more common types of such anomalies.[4]

When the aorta arches to the right, the descending aorta may lie either to the right or to the left of the spinal column. A right aortic arch means that the aorta arches to the right; it usually continues to descend upon the right. This anomaly produces a distinctive change in the course of the esophagus, which is discussed in Section A.

When the aorta arches to the right and is drawn back abruptly to the left and descends upon the left, the condition is described as a right aortic arch with a left descending aorta. In rare instances the reverse occurs: the aorta arches in the normal manner to the left and is drawn across to the right and descends to the right of the spinal column. Under such circumstances there is a left aortic arch and a right descending aorta.

In a number of instances both the left and right aortic arches persist and unite posteriorly to form the descending aorta; when this occurs the aorta encircles the trachea and the esophagus as a ring. A vascular ring is also formed when a right aortic arch is associated with a left descending aorta and the ductus arteriosus extends from the pulmonary artery to the descending aorta. Under such circumstances the aorta, the ductus arteriosus, and the pulmonary artery together form a ring which encircles the trachea and the esophagus. Section B

branchial arch distal to the pulmonary artery persists as the ductus arteriosus and the corresponding portion of the sixth left branchial arch atrophies and disappears Eventually the ductus arteriosus undergoes normal obliteration

Figure XXVI-3 shows a heart with a right aortic arch The upper drawing illustrates the position of the *aorta* and the origin of the *great vessels relative* to the esophagus The lower drawing shows a posterior view of the descending aorta and its relation to the esophagus and the azygos veins

A right aortic arch occurs in approximately 25 per cent of all patients with a tetralogy of Fallot and it is also common in other malformations which cause persistent cyanosis, especially those in which the aorta arises in part or entirely from the right ventricle, as in a truncus arteriosus, in a *complete* transposition of the great vessels or in a single ventricle with pulmonary stenosis It is known to occur with the Eisenmenger complex The *author has seen one patient* with a valvular pulmonary stenosis and an intact ventricular septum in whom the aorta arched to the right and Jones[1] has operated on one patient with an isolated patency of the ductus arteriosus who had a right aortic arch with the ductus on the right Such instances are, however rare

COURSE OF THE CIRCULATION

A right aortic arch is a vascular abnormality it affects the course of the aorta but places no strain upon the heart Although the aorta swings to the right instead of to the left this does not fundamentally alter the circulation of the blood

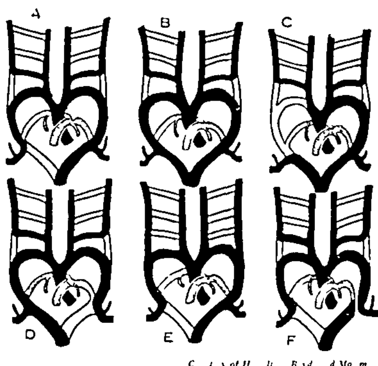
PHYSIOLOGY OF THE MALFORMATION

A right aortic arch in no way alters the physiology of the heart or the circulation

CLINICAL FINDINGS

The condition in itself is usually asymptomatic Only rarely does it alter th clinical picture as for example, if it occurs in combination with a coarctatic of the aorta in which the constriction is above the origin of the right subclavi artery Under such circumstances the left radial pulse will be stronger than t right

Hemiatrophy on the left is common in patients with a right aortic arch Tl is to be expected because a hemiatrophy is part and parcel of the underdevel ment of one side of the body With the alteration of stress and strain it is natu for the aortic arch to persist on the sturdier side



C i y of H li B yd d Mo m

FIGURE XXVI–2 Abnormalities in the development of the branchial arch arterial pattern

The persisting systemic arteries are represented in solid black the pulmonary trunk its branches and the ductus arteriosus are stippled (A) Normal pattern (B) double aortic arch (C) abnormal origin of the right subclavian artery (D) right aortic arch (E) absence of both carotid arteries (F) extreme degree of coarctation of the aorta the descending aorta being supplied by the ductus arteriosus alone

under discussion the fourth *left* branchial arch atrophies and the fourth *right* branchial arch persists to form the right aortic arch (see Figure XXVI–2D) The aorta may descend on the right of the spinal column throughout its entire length, or it may cross over in the lower thoracic region and emerge in its normal position beneath the diaphragm

NATURE OF THE MALFORMATION

When the aorta is developed from the fourth right aortic arch, the aorta arches to the right and descends upon the right Under such circumstances the three main vessels which arise from the arch of the aorta are the mirror image of the normal the first to be given off is the left innominate artery, then the right common carotid artery, and finally the right subclavian artery The sixth right

Stridor and *paralysis of the right vocal cord* have been reported in association with great dilatation of a right aortic arch in which the right recurrent laryngeal nerve passed beneath the arch of the aorta [5] This complication is extremely rare

CARDIAC FINDINGS

The cardiac findings are related to the associated malformation of the heart The course of the aorta is determined by fluoroscopy or x ray

X RAY AND FLUOROSCOPIC FINDINGS

A right aortic arch although asymptomatic, is readily detected upon x ray examination It should be sought for routinely in every cardiac fluoroscopy

The most significant positive finding is the visualization of the aortic knob to the right of the sternum When the aorta arches to the right, it pushes the superior vena cava abnormally far to the right This causes a broad ribbon like shadow to the right of the sternum which extends upward from the upper border of the heart to the clavicles The denser shadow of the aortic knob lies within the shadow cast by the superior vena cava It may be difficult to see the aortic knob upon fluoroscopy but it is usually readily discernible in the x ray, as shown in Figures xxvi-4 and 5

Visualization of the course of the esophagus clinches the diagnosis When the aorta swings to the right instead of the left it arches posteriorly on the right side of the esophagus It follows that the esophagus lies to the left of the aorta The relation of the normal aorta to the trachea and the esophagus is shown in Figure xxvi-6 and the relation of a right aortic arch to the same structures in Figure xxvi-7 (Figures xxvi-6 and 7 are shown in color on page 762)

In infancy the aortic knob is seldom visible Nevertheless in the anterior posterior position upon the administration of a radio-opaque mixture the esophagus normally is seen to lie in the mid line behind the shadow cast by the great vessels When the aorta arches to the right, it displaces the esophagus to the left and causes the esophagus to lie at the extreme left margin of the shadow at the base of the heart The position of the esophagus relative to the normal aorta and to a right aortic arch is shown in Figure xxvi-8 In infancy this single observation establishes the diagnosis In children and adults the relation of the aorta to the esophagus in the oblique positions is also of diagnostic aid In infants, however, the aorta is too small to produce any significant changes in the oblique positions

As the individual grows the aorta increases in size consequently the arch of the aorta presses upon the esophagus The impingement of the aorta upon the

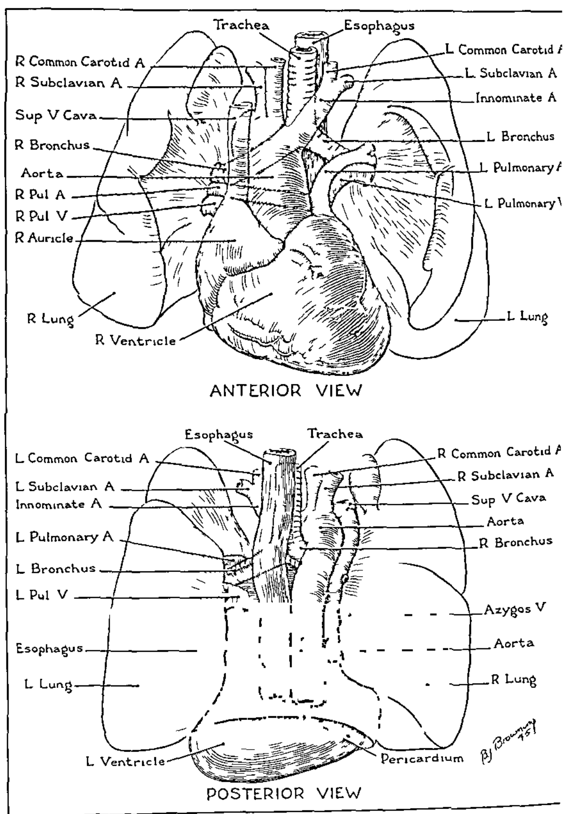


FIGURE XXVI-3 Tetralogy of Fallot with a right aortic arch

Stridor and *paralysis of the right vocal cord* have been reported in association with great dilatation of a right aortic arch in which the right recurrent laryngeal nerve passed beneath the arch of the aorta This complication is extremely rare

CARDIAC FINDINGS

The cardiac findings are related to the associated malformation of the heart The course of the aorta is determined by fluoroscopy or x ray

X RAY AND FLUOROSCOPIC FINDINGS

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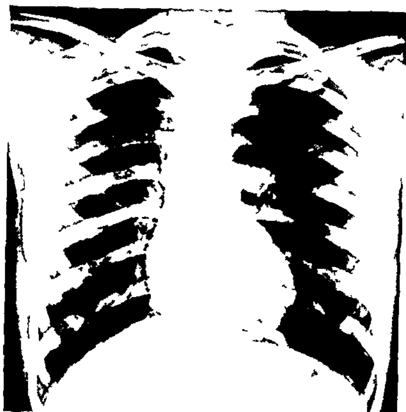
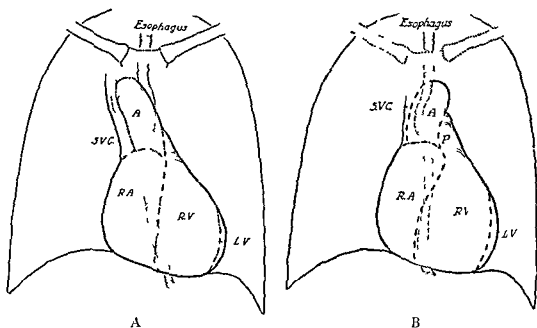


FIGURE XXVI–4 Tetralogy of Fallot with a right aortic arch Adult



A B

FIGURE XXVI–5 (A) Tetralogy of Fallot with a right aortic arch and (B) normal heart Adult

NOTE Figures XXVI–6 and 7 are shown in color on page 76-

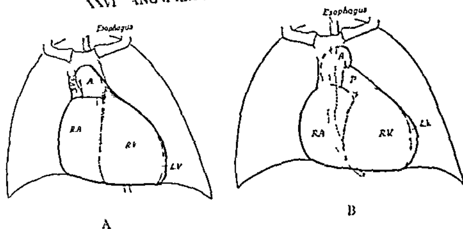


FIGURE XXVI-8 (A) Right aortic arch and (B) normal heart Infant

esophagus is visible both in the anterior posterior and in the oblique positions In the anterior posterior position the normal aorta impinges upon the left margin of the esophagus and indents it to the right When, however, the aorta arches to the right it impinges upon the right side of the esophagus and indents it to the left (see Figures XXVI-4 and 5)

In children and adults, the relation of the esophagus to the aorta is even more clearly seen in the oblique positions Normally it is in the right anterior-oblique position that the aorta is seen to impinge upon the esophagus and slightly displace it posteriorly, whereas in the left anterior-oblique position the aorta is not in contact with the esophagus If the aorta arches to the right instead of to the left, the esophagus is independent of the aorta in the right anterior-oblique position, in the left anterior-oblique position the aorta may or may not press upon the esophagus and indent it posteriorly (Figures XXVI-9 and 10) The two oblique positions are not the exact mirror image of the normal, because, although the aortic arch is the mirror image of the normal the heart itself occupies its normal position

DIAGNOSIS

The diagnosis is based upon the x ray and fluoroscopic findings It depends on simple awareness of the possibility of the condition and accuracy of observation For this reason whenever an esophagram is obtained special attention should be paid to the relation of the esophagus to the aorta Inasmuch as the anomaly occurs most frequently in association with a partial transposition of the

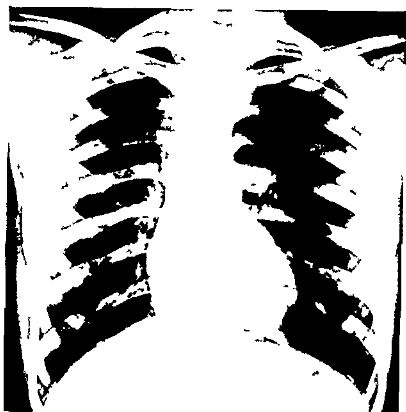


FIGURE XXVI-4 Tetralogy of Fallot with a right aortic arch Adult

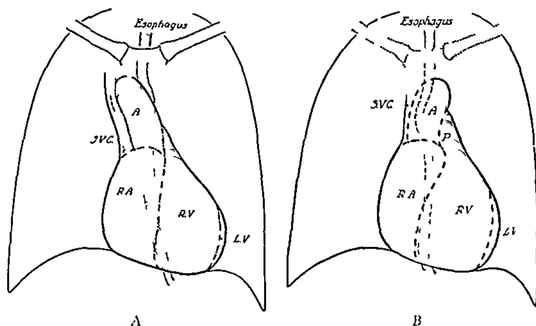
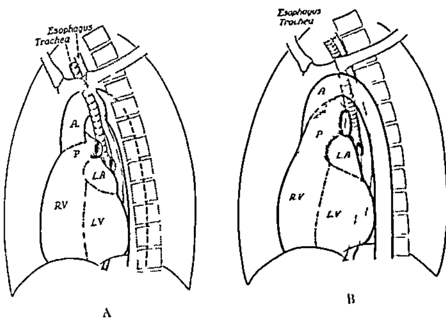
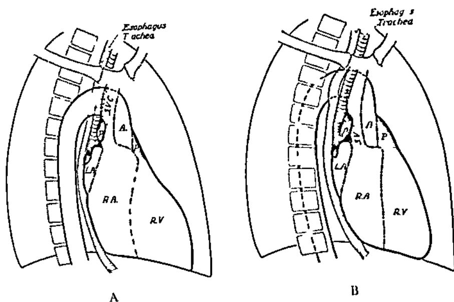


FIGURE XXVI-5 (A) Tetralogy of Fallot with a right aortic arch and (B) normal heart Adult

NOTE Figures XXVI-6 and 7 are shown in color on page 76[illegible]



LEFT ANTERIOR-OBLIQUE POSITION



RIGHT ANTERIOR-OBLIQUE POSITION

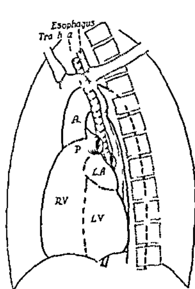
FIGURE XXVI–10 (A) Tetralogy of Fallot with a right aortic arch and (B) normal heart Adult

Left
anterior-oblique
position

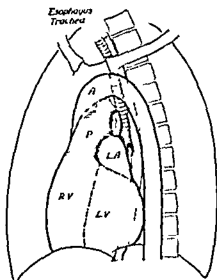
Right
anterior oblique
position



FIGURE XXVI-9 Tetralogy of Fallot with a right aortic arch Adult

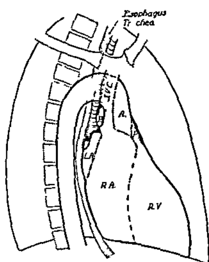


A

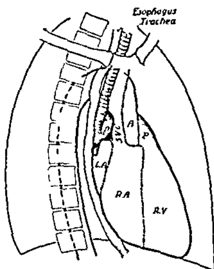


B

LEFT ANTERIOR-OBLIQUE POSITION



A



B

RIGHT ANTERIOR-OBLIQUE POSITION

FIGURE XXVI–10 (A) Tetralogy of Fallot with a right aortic arch and (B) normal heart Adult

great vessels, the relation of the esophagus to the aorta should always be studied with great care in all patients who show persistent cyanosis

DIFFERENTIAL DIAGNOSIS

The condition may call for differentiation from *aneurysmal dilatation of the ascending aorta* Such dilatation may occur as an isolated anomaly or in association with some malformation of the heart *Coarctation of the aorta* may be confused with a right aortic arch because of the absence of the aortic knob and the dilatation of the ascending aorta Visualization of the course of the esophagus should immediately clarify the nature of the anomaly

PROGNOSIS

The prognosis is excellent The malformation in itself has no effect upon the life expectancy of the individual The anomaly is of clinical importance only in those malformations, amenable to surgery, in which the operation concerns the aorta or the vessels which arise therefrom

SUMMARY

A right aortic arch is the result of the persistence of the fourth right branchial arch instead of the fourth left branchial arch The anomaly occurs in approximately 25 per cent of all patients with a tetralogy of Fallot It also occurs in other malformations in which the aorta arises in part or entirely from the right ventricle It is the rule for the aortic arch to persist on the right when there is a left hemiatrophy In rare instances a right aortic arch may occur as an isolated malformation

The condition is asymptomatic

The diagnosis is made by x ray or fluoroscopy The aortic knob lies to the right of the sternum within the shadow cast by the superior vena cava

An esophagram readily confirms the diagnosis In infants, although the aortic knob may not be clearly visible, the esophagus is seen to lie at the extreme left margin of the shadow cast by the great vessels In children and adults, as the aorta arches across the esophagus, it impinges upon the right margin of the esophagus and indents it to the left Examination in the right anterior oblique position confirms the presence of a right aortic arch, as the course of the esophagus is entirely independent of the aorta

The condition occasionally calls for differentiation from aneurysmal dilatation of the ascending aorta and from coarctation of the aorta

The prognosis is excellent The condition is of importance only in relation to cardiac surgery, especially a Blalock Taussig operation

B *Vascular Rings*

When the aorta arches to the right and is drawn abruptly back to *the left and* descends upon the left, the condition is known as a right aortic arch with a left descending aorta Under such circumstances the aorta lies behind the esophagus thus, the aorta is *retro-esophageal* The relation of the aorta to the esophagus is shown in Figures XXVI-11 and 12 Bedford and Parkinson *reported three condi* tions which, in combination with a right aortic arch, cause the aorta to descend

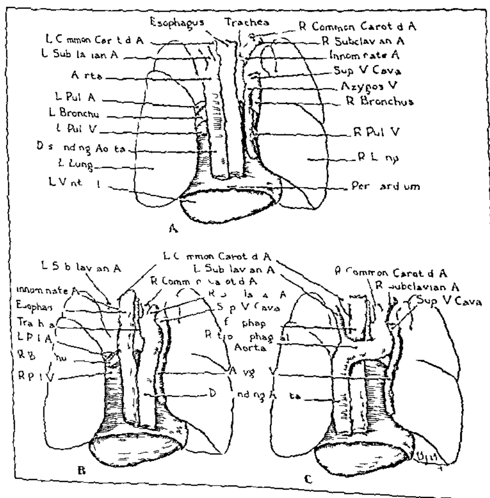


FIGURE XXVI-11 (A) Normal heart (B) right aortic arch (C) right aortic arch and left descending aorta Posterior view

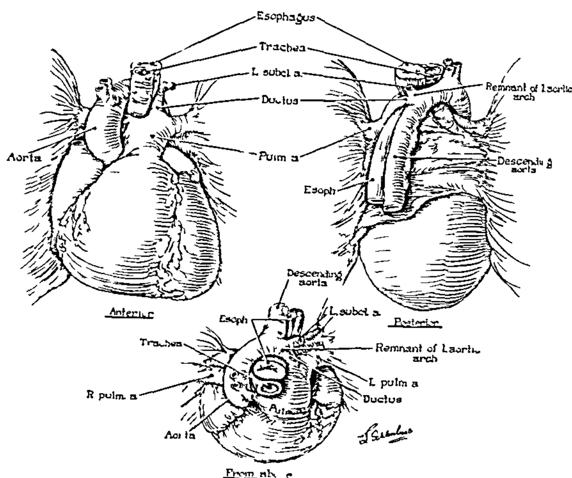


FIGURE XXVI–12 Double aortic arch or aortic ring

on the left The first is the normal development of the ductus arteriosus on the left side or the persistence of its strand in its normal position, the second is the origin of the left subclavian artery from the left aortic root the third is the development of the right aortic arch and the persistence of the left aortic arch the result being a vascular ring which encircles the trachea and the esophagus The third condition is known as a double aortic arch, an aortic ring or a vascular ring

A fourth variation was reported by Paul [6] namely, a left aortic arch and right descending aorta Under such circumstances the aorta arches to the left in the normal manner and is drawn back to the right and descends upon the right This condition also causes a retro esophageal aorta

By far the most important of these conditions is the vascular ring When the great vessels encircle the trachea and esophagus, the resultant ring may be so small as to cause serious constriction The symptoms are caused by the constric

tion of the trachea and the esophagus The signs are caused by their displacement

A retro-esophageal aorta without a complete vascular ring may cause symptoms if the aorta compresses and displaces the trachea and esophagus anteriorly as it courses posteriorly to these vessels If such is the case, the symptoms are identical with those produced by a vascular ring Therefore these anomalies are considered together

NATURE OF THE MALFORMATION

A vascular ring is formed when a right aortic arch occurs in combination with a left descending aorta and the ductus arteriosus develops in the normal manner from the sixth left branchial arch Under such circumstances the ductus arterio us, or after birth a strand thereof draws the aorta sharply to the left, posterior to the trachea and esophagus, the aorta descends in the normal manner to the left of the spinal column Thus the aorta, the pulmonary artery, and the ductus arteriosus or the fibrous strand of the obliterated ductus form a ring around the trachea and esophagus (see Figure XXVI–13) This is the commonest type of a vascular ring and frequently occurs with an otherwise normal heart and vascular system

Although extremely rare, the corresponding anomaly on the right may occur that is a left aortic arch a right descending aorta and a ductus arteriosus, formed from the sixth right branchial arch, may persist on the right and extend from the pulmonary artery to the right descending aorta In this manner, also, the trachea and esophagus may be encircled (Figure XXVI–14)

A double aortic arch means that both the right and left aortic arches persist as functioning tubes and unite posteriorly with the dorsal aorta This, too, forms a vascular ring which encircles the trachea and esophagus The descending aorta may lie on the right or on the left That on the left is, however normal hence such is the more common location Although both aortic arches may carry blood when there is a double aortic arch the right aortic arch is usually but not always the larger of the two Arkin[7] was the first to report this condition Figure XXVI–15 illustrates the relation of a double aortic arch or aortic ring to the trachea and the esophagus

Griswold and Young have reviewed the various types of vascular rings and added two cases of double aortic arch In one patient the right component of the ring was the smaller of the two and in the other the left was the smaller It is worthy of note that with the persistence of a right aortic arch, a portion of

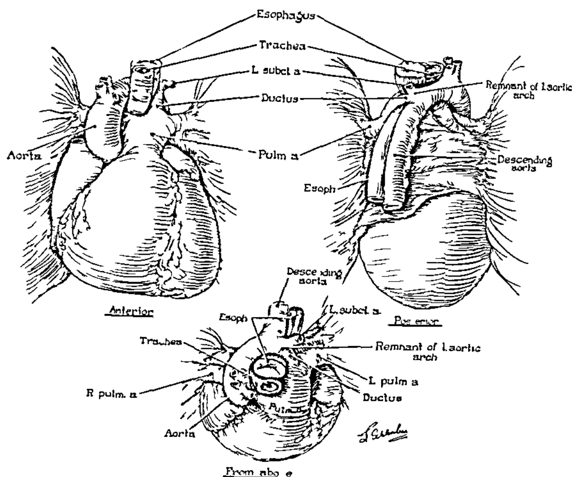


FIGURE XXVI 12 Double aortic arch or aortic ring

on the left The first is the normal development of the ductus arteriosus on the left side or the persistence of its strand in its normal position, the second is the origin of the left subclavian artery from the left aortic root, the third is the development of the right aortic arch and the persistence of the left aortic arch the result being a vascular ring which encircles the trachea and the esophagus The third condition is known as a double aortic arch, an aortic ring, or a vascular ring

A fourth variation was reported by Paul [6] namely a left aortic arch and right descending aorta Under such circumstances the aorta arches to the left in the normal manner and is drawn back to the right and descends upon the right This condition also causes a retro esophageal aorta

By far the most important of these conditions is the vascular ring When the great vessels encircle the trachea and esophagus, the resultant ring may be so small as to cause serious constriction The symptoms are caused by the constric

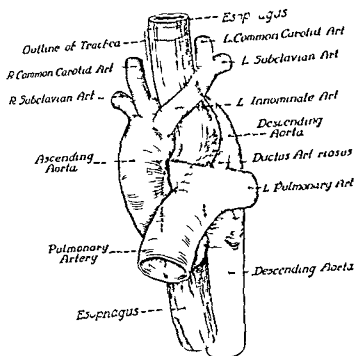


FIGURE XXVI 13 Right aortic arch with the descending aorta drawn to the left by the ductus arteriosus

the left segment may be partially or completely obliterated and only a strand of tissue remain when, however, the right component persists, it always carries blood Nevertheless the left arch may be the larger of the two and under such circumstances an operation for the relief of symptoms is best performed upon the right side Therefore it is desirable, when possible to determine which of the two components is the larger that is, which is the main component

The symptoms are caused by the displacement and compression of the trachea and esophagus by the retro-esophageal aorta Even when there is no vascular ring if the subclavian artery arises from the descending aorta and draws the aorta back to the opposite side from the direction of the aortic arch, the displacement of the aorta may be so great as to cause the same signs and symptoms as those produced by a vascular ring Hence all these conditions are considered together

Finally it is important to remember that although the aorta usually lies behind both the esophagus and the trachea in rare instances the aorta may lie between these vessels

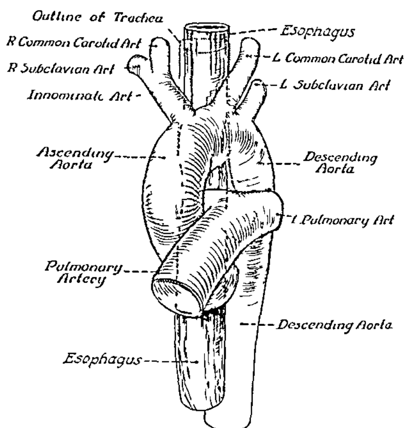


FIGURE XXVI-6 Relation of a normal aorta to the trachea and esophagus

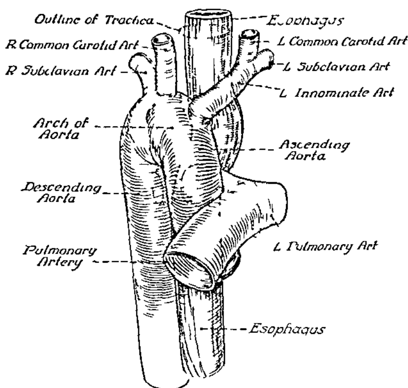


FIGURE XXVI-7 Relation of a right aortic arch to the trachea and esophagus

the aorta after it has arched to the right, to be drawn to the left It follows that instead of arching posteriorly and descending on the right, the arch of the aorta occupies a transverse position and turns abruptly downward at the level of the left subclavian artery Consequently the arch of the aorta crosses from right to left behind the esophagus, that is, between the spinal column and the esophagus When it is recalled that the arteries are able to erode bones, it is not surprising to find that the aorta is so much more powerful than the trachea and the esophagus that, when the aorta is drawn abruptly to the left, the retro-esophageal aorta causes an anterior displacement of the trachea and the esophagus

When there is a normal left aortic arch and a right descending aorta,[8] the condition is the mirror image of a right aortic arch with a left descending aorta The same possible variations exist that is, the ductus arteriosus fully develops from the sixth right branchial arch and persists on the right, or the right subclavian artery may arise from the descending aorta

All these anomalies involve the development of the aortic arch and its relation to the esophagus In each instance the anomaly concerns the arch of the aorta the transverse arch lies posterior to the esophagus and the descending aorta lies on the opposite side to that of the ascending aorta Thereafter the anomaly ceases and the remainder of the vascular system follows its normal pattern This may be the reason why the apparently more extreme anomaly of the double aortic arch frequently occurs as an isolated abnormality in an otherwise normal heart

COURSE OF THE CIRCULATION

Although the aorta follows an abnormal course, the flow of blood throughout the body is unimpeded The course of the circulation is normal The anomaly places no strain upon the heart or the circulation

PHYSIOLOGY OF THE MALFORMATION

The physiology is altered only by the compression of the trachea and the esophagus

CLINICAL FINDINGS

The symptoms are produced by the constriction of the trachea and the esophagus hence they are the same in the various types of vascular rings When the constriction is severe the signs and symptoms date from birth or early infancy

Attacks of croup and *pulmonary infections* are common in infancy and early

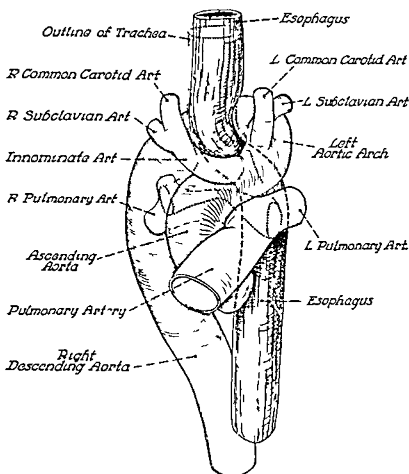


FIGURE XXVI–14 Left aortic arch and right descending aorta

When the aorta arches to the right and the left subclavian artery arises from the dorsal root of the aorta, the resultant distortion is similar to that which occurs when the ductus arteriosus is on the left Under such circumstances, even though the aorta arches to the right, the left subclavian artery follows the normal pattern and arises from a persistent left aortic root which, in turn, takes its origin from the left dorsal aorta This causes the transverse arch of the aorta to be drawn abruptly to the left, posterior to the esophagus, leaving the aorta to descend upon the left in the normal manner Figure XXVI–16 illustrates the manner in which the aorta ascends on the right, is drawn back to the left behind the trachea and the esophagus by the left subclavian artery, and descends upon the left In such cases either the right innominate artery, the right subclavian artery, or the left common carotid artery may be the first vessel to be given off from the arch of the aorta, the left subclavian artery, however, always arises from the descending aorta [9, 10] The abnormal origin of the left subclavian artery causes

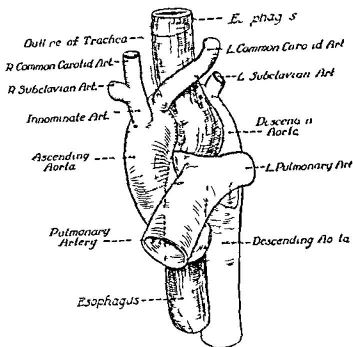


FIGURE XXVI-16 Right aortic arch with the descending aorta drawn to the left by the left subclavian artery which arises from the dorsal root of the aorta

Attacks of paroxysmal dyspnea are not uncommon The attack may be precipitated by food or by the swallowing of saliva, or it may occur without any apparent cause

The infant becomes extremely dyspneic and may become intensely cyanotic Indeed the infant may die of asphyxia These attacks can usually be relieved by hyperextension of the head and neck

Aspiration pneumonia is a real and ever present danger Many of these infants die from this cause

Undernutrition may be conspicuous Many of these infants eat so slowly and have so much difficulty swallowing that their nutrition suffers Nevertheless, it is important to emphasize that some infants grow and develop normally during the first months of life and have no difficulties until they are between nine and twelve months of age

The late development of symptoms toward the end of the first year of life, is in all probability caused by the contraction of the strand of fibrous tissue which is formed after the ductus arteriosus undergoes obliteration

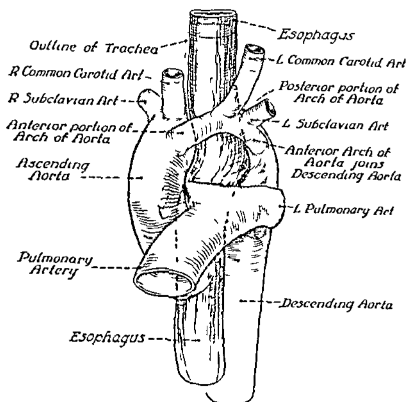


FIGURE XXXI-15 Double aortic arch or aortic ring

childhood The compression of the trachea by the retro esophageal aorta renders the infant extremely prone to *tracheitis laryngitis* and *bronchitis*

A brassy cough is of frequent occurrence, it is caused by the pressure upon the left recurrent laryngeal nerve Not infrequently the mother brings the infant to the physician with severe croup, a brassy cough, and congestion in the lungs It is the persistence of the brassy cough after the pulmonary infection has cleared which gives the clue to the diagnosis

Fortunately, as the child grows, the trachea increases in size and its compression becomes less serious hence symptoms are less marked in older patients than in infants

Stridor is also a common complaint

Retraction of the head with each inspiration may occur when the constriction is marked In addition, there may be marked retraction of the lower costal ribs with each breath

Dysphagia may be severe An infant with a vascular ring may be asymptomatic until solid foods are started For this reason, when there is a history of dysphagia, if barium is to be given it must be thin, as a thick barium mixture may precipitate a severe attack of choking and paroxysmal dyspnea

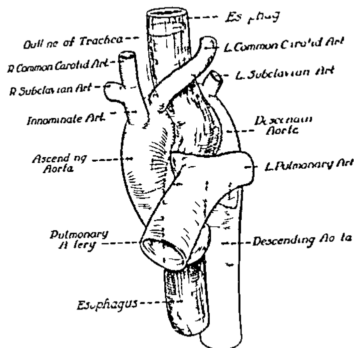


FIGURE XXVI-16 Right aortic arch with the descending aorta drawn to the left by the left subclavian artery which arises from the dorsal root of the aorta

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Aneurysms of the transverse arch of the aorta if they occur in combination with a retro esophageal aorta, are more serious than when the aorta is normally placed Since the aorta lies between the trachea and the spinal column, as the aorta dilates it may erode the spine and cause pain and serious pressure symptoms Such complications are comparatively rare and occur only in adults

CARDIAC FINDINGS

The cardiac manifestations are minimal In contrast to a simple right aortic arch, which occurs most commonly in association with malformations of the great vessels, the right aortic arch with a left descending aorta is usually an isolated anomaly in an otherwise normal cardiovascular system

X RAY AND FLUOROSCOPIC FINDINGS

The displacement of the esophagus causes a characteristic x ray picture When the aorta arches to the right and is drawn back to the left, posterior to the esophagus, *it displaces the esophagus anteriorly* Usually this displacement can be detected in the anterior posterior position by a kink in the esophagus (see Figures XXVI-17 and 18) The anterior displacement of the esophagus in the



FIGURE XXVI-17 Retro esophageal aorta Infant

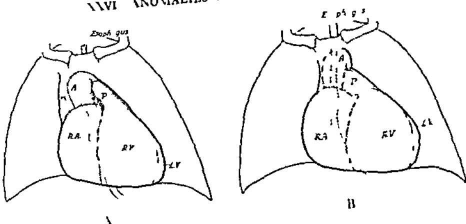


FIGURE XXVI-18 (A) Retro-esophageal aorta and (B) normal heart Infant

oblique views and in the lateral views is unmistakable The esophagus makes an abrupt curve anteriorly at the point where the arch of the aorta impinges upon it (see Figures XXVI-19 and 20) The area of the compression of the esophagus is approximately the size of the aorta

When the aorta arches to the left and is drawn abruptly over to the right and descends upon the right, the aorta lies posterior to the esophagus and displaces the esophagus anteriorly The aortic knob, however, is visible on the left and the esophagus is seen to lie to the extreme left of the cardiac shadow immediately below the aortic knob (see Figures XXVI-21 and 22) Such findings, combined with evidence of a retro-esophageal aorta in the oblique views (see Figures XXVI-23 and 24) were reported by Paul[6] to be characteristic of a left aortic arch and a right descending aorta

The course of the esophagus is identical in infancy and in adult life It is, however worthy of note that owing to the more transverse position of the heart, the abnormal course of the aorta produces a greater increase in the width of the shadow at the base of the heart in infants than in older individuals Indeed, in infancy a retro-esophageal aorta may cause an abnormally wide mediastinal shadow (see Figure XXVI-17)

DIAGNOSIS

Awareness of the possibility of this anomaly and observation of the course of the esophagus after the administration of a single teaspoonful of barium are all that are needed to establish the diagnosis It takes but a moment No other condition causes a similar anterior displacement of the esophagus

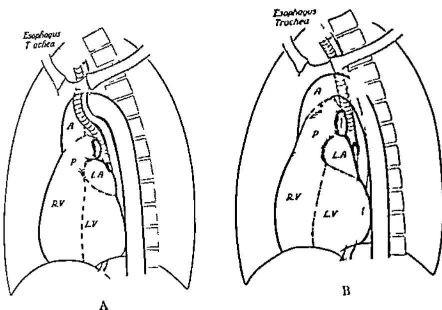


Left anterior-oblique position



Right anterior-oblique position

FIGURE XXVI-19 Right aortic arch with a left descending aorta Infant



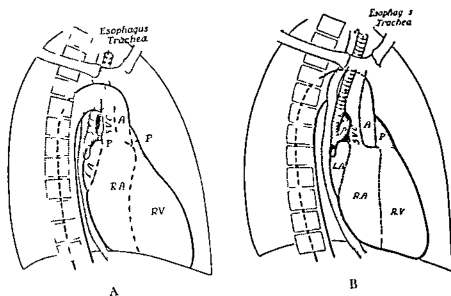


FIGURE XXVI-20 (A) Right aortic arch with a left descending aorta and (B) normal heart Infant



FIGURE XXVI–21 Left aortic arch with a right descending aorta Child

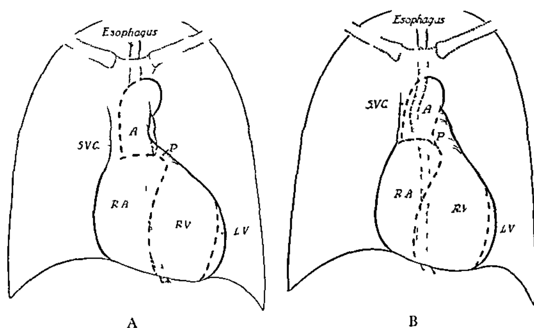


FIGURE XXVI–22 (A) Left aortic arch with a right descending aorta and (B) normal heart Adult

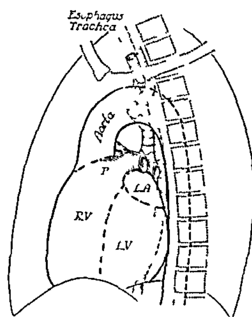


Left anterior-oblique position

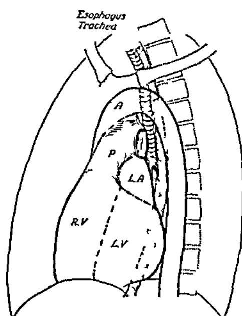


Right anterior-oblique position

FIGURE XXVI–23 Left aortic arch with a right descending aorta Child

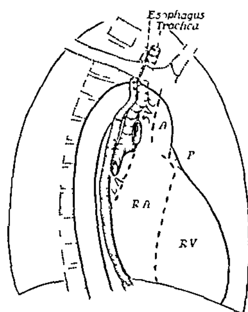


A

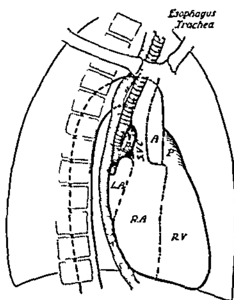


B

LEFT ANTERIOR-OBLIQUE POSITION



A



B

RIGHT ANTERIOR-OBLIQUE POSITION

FIGURE XXVI-24 (A) Left aortic arch with a right descending aorta and (B) normal heart Adult

It is essential to remember that for infants the barium mixture, if used at all, must be thin The mixture should be of the consistency of cream soup, because in the presence of a ring there is real danger *that the infant may* choke and even suffocate to death if the barium mixture is thick and sticky Indeed, when the symptoms suggest the presence of a vascular ring, barium should not be used, because of the danger of both aspiration and suffocation For this reason, lipiodol or some other substance which is not harmful to the lungs and which is readily swallowed, should be used for the delineation of the esophagus

Arkin[7] and Gross[11] have both emphasized that, in the presence of an aortic ring the esophagus is constricted anteriorly as well as on its posterior surface by the two components of the ring This is well illustrated in the lower x ray of Figure XXVI–23

It is important to remember that *in rare instances the aorta may pass between* the esophagus and the trachea Therefore if the infant is suffering from symptoms suggestive of a vascular ring and delineation of the esophagus fails to reveal evidence of a retro-esophageal aorta, the possibility remains that the aorta passes behind the trachea Under such circumstances delineation of the course of the trachea with lipiodol may reveal the anterior displacement of the trachea

DIFFERENTIAL DIAGNOSIS

The differentiation of a right aortic arch combined with a left descending aorta caused by the persistence of a strand of the ductus arteriosus, or even of the ductus itself, from the distortion of the arch due to the anomalous origin of the left subclavian artery, or even from a double aortic arch, is extremely difficult and, indeed, may be impossible Even with angiocardiography or aortography one gets a two-dimensional image of a three-dimensional object Hence it may not be possible to distinguish the structure of the ring The condition is further complicated by the fact that a portion of the ring may not be a blood-containing vessel The important consideration is not the anatomical structure of the vascular ring, but its differentiation from other retro-esophageal vessels and from other conditions

A retro esophageal aorta without a vascular ring seldom causes symptoms The anterior displacement of the aorta is in the same location and may be quite as marked A brassy cough may occur because of pressure upon the recurrent laryngeal nerve but dysphagia and retraction of the head and chest with each inspiration are seldom seen The condition is usually asymptomatic and picked up on fluoroscopic examination If symptoms develop in adult life, other causes

should be sought for The author has known one adult with this condition whose symptoms of dysphagia proved to be an early sign of scleroderma In another patient the dysphagia was due to a globus hystericus

A retro esophageal subclavian artery causes similar anterior displacement of the esophagus but the indentation of the esophagus is smaller, corresponding to the smaller size of this vessel The condition is usually asymptomatic It may, however, cause symptoms Indeed, a right retro-esophageal subclavian artery was mentioned by Bedford and Parkinson[5] as a cause of *dysphagia lusoria* If the condition does lead to difficulty, the abnormality is amenable to surgery (see below)

A retro esophageal innominate artery is rare, when it does occur, it causes an indentation of the esophagus corresponding to the size of that vessel

Bronchial arteries and anomalous vessels of collateral circulation may cause anterior displacement of the aorta at almost any level, but usually these vessels pass behind the esophagus in the lower thoracic region Occasionally the esophagus is caught between these vessels and distorted in a bizarre manner as previously shown in Figure xiv–19

An aortic pouch or diverticulum may occur when the aorta arches to the right and descends upon the right Under such circumstances, even though the left subclavian artery may arise from the anterior arch of the aorta, there may be a pouching of the aorta at the level of the left aortic root The diverticulum lies behind the trachea and esophagus and causes displacement of the esophagus which may be visible in the anterior posterior position as well as in the oblique views The anterior displacement of the esophagus, however, lies at a lower level than does the arch of the aorta It is the location of the anterior displacement of the aorta which aids in the differentiation of this condition from a true vascular ring

In infants the condition also calls for differentiation from other causes of tracheal obstruction and of wide mediastinal shadows In adults the condition may be mistaken for coarctation of the aorta, substernal thyroid, or other mediastinal tumors or an aneurysm of the arch of the aorta

A croupy cough and *repeated pulmonary infections* direct attention to the lungs rather than to the heart Observation of the course of the esophagus after the administration of a teaspoonful of barium immediately reveals the nature of the difficulty

A wide mediastinal shadow always brings to mind the possibility of a persistent thymus In the left anterior oblique position it is usually possible to visual

ize the outline of the thymus anterior to the cardiac shadow The thymus often causes a wing like projection of the shadow just above the upper margin of the cardiac silhouette (see Figure 11-13) The differentiation of a *persistent thymus* from the malformation under discussion is simple, *provided the course of the esophagus* is delineated by barium

Coarctation of the aorta resembles the anomaly under discussion in that the aortic knob is absent and there is dilatation of the ascending aorta The absence *of other signs of coarctation* and the delineation of the course of the esophagus readily differentiate the two conditions

Substernal thyroids and other masses in the anterior mediastinum do *not* cause anterior displacement of the esophagus

Aneurysm of the transverse arch of the aorta may produce neurological symptoms by pressure on the spinal cord Again, awareness of the possibility and observation of the course of the esophagus after the administration of barium permit accurate diagnosis

TREATMENT

Attacks of paroxysmal dyspnea, when due to a vascular ring, call for hyper extension of the head and neck Such attacks may cause the infant to become cyanotic, nevertheless, the dyspnea always precedes the appearance of cyanosis and between attacks the infant s color is normal

Surgery is required to eliminate the underlying abnormality Gross[11] was the first to show that the constriction of the trachea and esophagus could be relieved by surgery Indeed, if the infant is suffering from attacks of paroxysmal dyspnea and cyanosis, the condition is almost a surgical emergency It seems as though the ductus arteriosus, in the process of obliteration, clamps down spasmodically and thus causes the vascular ring to contract Such infants are in real danger of dying of asphyxia Surgery may save the infant s life and restore the circulation to normal

If the vascular ring is caused by a right aortic arch and the strand of the duc tus arteriosus which persists on the left and draws the aorta to the left behind the esophagus, division of the ductus arteriosus is indicated

The entire length of the ductus arteriosus should be dissected away from the esophagus This is important because frequently its distal portion is so forcibly bound by adhesions to the esophagus that it continues to compress and constrict the esophagus even though it has been divided Gross has emphasized the im portance of suturing the anterior mediastinum to the sternum in an effort to re

lieve still further the compresssion of the trachea and esophagus, because, although theoretically division of the ring cures the condition, frequently the distortion and constriction of the esophagus persist after surgery

If the symptoms are due to a double aortic arch, division of the smaller segment is indicated It is, however, essential to be certain that the remaining segment is patent throughout its course Inasmuch as the left segment may be partially obliterated and such an obliteration seldom, if ever, occurs on the right, it is usually wiser to operate on the left side

In rare instances the distortion of the aorta and the symptoms caused thereby are due to the anomalous origin of the left subclavian artery from the descending aorta Under such circumstances, simple division of this artery will relieve the pressure of the aorta upon the trachea and esophagus Since Dr Alfred Blalock and his associates have sacrificed the subclavian artery in over 1,500 patients without injury to the arm, the risk of such a procedure is minimal

A retro-esophageal aorta or a vascular ring may produce no symptoms throughout life Indeed, if the patient is asymptomatic and over two years of age, there is no reason to believe that he will develop difficulty Under such circumstances there is no necessity for surgery

PROGNOSIS

An infant in whom a vascular ring causes severe symptoms usually dies before one year of age unless the condition is relieved by surgery For such an infant surgery is indicated and the prognosis is guarded

If, however, the symptoms are mild, conservative therapy is indicated, because as the infant grows the trachea becomes stronger, the pressure on the trachea becomes proportionally less, and the symptoms may completely disappear

Patients who are asymptomatic in infancy remain so throughout their lives unless the condition is later complicated by acquired vascular diseases The anomalies in themselves are readily compatible with life and need cause no concern

SUMMARY

A vascular ring is formed when there is a right aortic arch and a left descending aorta and the ductus arteriosus or a strand thereof persists on the left A vascular ring is also formed when the aorta arches to the left but descends upon the right and the ductus arteriosus or a strand thereof persists on the right An aortic

ring or a double aortic arch occurs when both the right and left aortic arches persist and join posteriorly to form the descending aorta A retro-esophageal aorta also occurs when the aortic arch develops on one side and the opposite subclavian artery develops anomalously from the root of the dorsal aorta, thus drawing the descending aorta behind the trachea and the *esophagus* and causing the aorta to descend upon the opposite side from that of the arch of the aorta Such anomalies usually occur as isolated abnormalities in an otherwise normal cardiovascular system

The condition, when extreme, constricts the trachea and the esophagus and may cause serious difficulty in early infancy

The outstanding symptoms are attacks of croup, a brassy cough, stridor, and severe attacks of paroxysmal dyspnea Dysphagia may be severe Nevertheless, it may not develop until the infant is started on solid food An attack of paroxysmal dyspnea may be fatal

The cardiac findings are normal

The diagnosis is based upon the x ray and fluoroscopic findings The aortic knob may be visible in the anterior posterior view on the right or the left In the oblique views and often in both lateral views, there is a sharp anterior displacement of the trachea and the esophagus at the level of the aortic arch It is produced by the aorta as it passes behind these structures No other condition causes a similar marked displacement

The condition may require differentiation from smaller vessels which pass behind the trachea and the esophagus and also from other causes of cough and tracheal obstruction and from a substernal thyroid, a mediastinal *tumor*, and an aneurysm of the arch of the aorta

The immediate treatment of an attack of paroxysmal dyspnea is hyperextension of the head and neck The occurrence of such attacks is usually an indication for immediate surgery

Surgery is necessary to relieve the underlying constriction caused by the vascular ring It is usually best performed on the left side It is important to free all the tissues as much as possible and to dissect the entire length of the ductus and also to suture the anterior mediastinum to the sternum in order more effectively to relieve the constriction of the trachea and the esophagus

The prognosis in infants with symptoms is serious, but it may be completely altered by surgery If the patient is asymptomatic for the *first two* years of life, the condition will probably never cause difficulty Under such circumstances surgery is contraindicated

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drawn to the author's attention by an illustration in a Russian atlas on congenital malformations of the heart,[13] in this instance the two ascending aortae arose from the left ventricle From each ascending aorta, three great vessels arose the innominate, the common carotid, and the subclavian artery The two aortae encircled the trachea and the esophagus and united posteriorly to form a common descending aorta The first such case is said to have been reported by Malacarne in 1788 * The author was told that, in Malacarne s case and also in the three cases kindly located for her by Dr W C Manion at the Armed Forces Institute of Pathology in Washington, there were two superior venae cavae and a single inferior vena cava In the specimen shown in Figure XXVI-25 one aorta arises from the left ventricle and the other aorta arises from the right ventricle, which gives the right ventricle the appearance of a single ventricle In the Russian illustration the two aortae arise from the left ventricle When in the U S S R in 1958, the author inquired from Dr A A Vishnevsky concerning this anomaly but found that there was no such specimen in their collection and that they were unable to give her any details about the anomaly Dr W C Manion[14] advised the author that in Malacarne s original report and in all three specimens in the Armed Forces Institute of Pathology, this anomaly occurred in a Siamese monster with two heads and a single body!

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5 Bedford D E and J Parkinson Right sided aortic arch (*situs inversus arcus aortae*) Brit J Radiol 9 776–798 1936

6 Paul R N A new anomaly of the aorta—left aortic arch with right descending aorta J Pediat 32 19–29 1948

7 Arkin A Double aortic arch with total persistence of the right and isthmus stenosis of the left arch a new clinical and x ray picture report of 6 cases in adults Am Heart J 11 444–474 1936 (First case reported in Wien Arch inn Med 12 385–416, 1926)

* The author has been unable to locate the original article but did find a report by Malacarne of a man of sixty years who had a double aortic arch with what was described as two ascending aortae this was published in Malacarne s *Delle osservazioni in chirurgia* (Turin G M Briolo 1784)

C *Other Anomalies of the Aortic Arch*

There are many variations in the origin of the great vessels as they arise from the aortic arch The vast *majority of* these variations are entirely asymptomatic but may be *of clinical* importance in the surgical *correction of* malformations of the heart

VARIATIONS IN THE ORIGIN OF THE GREAT VESSELS

Anomalies of the great vessels are extremely numerous There *may be no* innominate artery and four *great vessels* The *right* subclavian artery may arise as *a branch* of the left subclavian artery[8, 12] Both common carotid arteries may *arise as* branches from the innominate artery All four vessels may arise *from* a common trunk, as they normally do in the horse

Dr Osler Abbott has told the author of a patient with a right aortic arch in *whom* there were only two great vessels arising from the arch of the aorta one was the right subclavian artery and the other was a large *trunk which ran be tween the* trachea and the esophagus and gave rise to the right common carotid artery, then the left common carotid artery, and finally the left subclavian ar tery The ligament of the obliterated ductus arteriosus was given off below the level of the left common carotid *artery This arrangement* of the great vessels has *also* been found in two children with tracheo-esophageal fistula Figure xxvi–2 *gives* a schematic representation of the common anomalies in the development of the great vessels

Variations in the origin of the great vessels are common in a tetralogy of Fallot even when the aorta follows its normal course, that is, when it arches to the left and descends upon the left As previously mentioned, when the aorta arches *to the right,* the great vessels should be the mirror image of the normal (see Section A) Nevertheless, variations in the position of the great vessels are quite as common when the aorta arches to the right as they are in a tetralogy of Fallot with a left aortic arch

When a *dextrocardia* is of such a nature as to cause persistent cyanosis or when there is a situs inversus, the aorta may descend on the right or on the left A right aortic arch is, of course, normal when the primitive cardiac tube swings to the right instead of to the left Nevertheless, a left aortic arch occurs more *frequently* in a dextrocardia with persistent cyanosis than does a right aortic arch with a tetralogy of Fallot (see Chapter xxxiii)

TWO ASCENDING AORTAE

A vascular ring with two ascending aortae is a rare anomaly which was first

8 *Griswold H E Jr* and M D Young Double aortic arch, report of 2 cases and review of the literature Pediatrics 4 751–768, 1949

9 Abbott M E Atlas of Congenital Cardiac Disease New York American Heart Association 1936

10 Dolgopol V B Anomalous origin of the right subclavian artery from the descending arch of aorta J Tech Methods 13 112–118 1934

11 Gross R E Surgical relief for tracheal obstruction from a vascular ring New England J Med 233 586–590 1945

12 Roche U J I Steinberg and G P Robb Right sided aorta with descending aorta simulating aneurysm Arch Int Med 67 995–1007 1941

13 Daschagaran A D Atlas on Congenital Malformations of the Heart Moscow Dr A A Vishnevsky Clinic 1957

14 Manion W C Personal communication

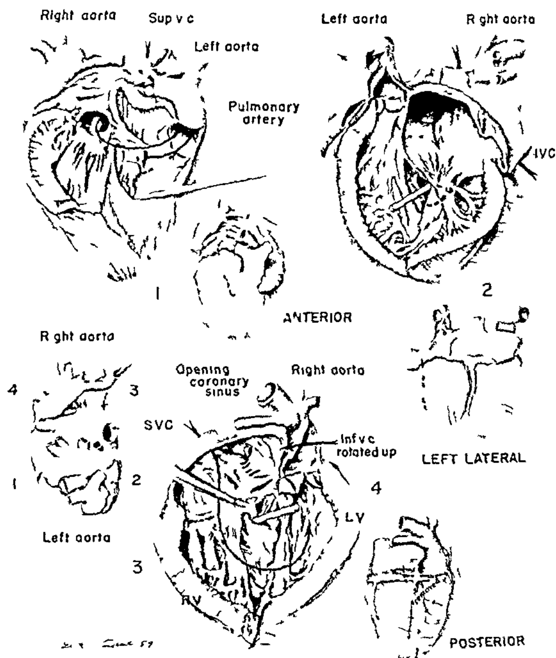


Figure XXVI 25 Vascular ring with two ascending aortae

8 Griswold H E Jr and M D Young Double aortic arch, report of 2 cases and review of the literature Pediatrics 4 751–768 1949
9 Abbott M E Atlas of Congenital Cardiac Disease New York American Heart Association 1936
10 Dolgopol V B Anomalous origin of the right subclavian artery from the descending arch of aorta J Tech Methods 13 112–118 1934
11 Gross R E Surgical relief for tracheal obstruction from a vascular ring New England J Med 233 586–590 1945
1 Roche, U J, I Steinberg and G P Robb Right sided aorta with descending aorta simulating aneurysm Arch Int Med 67 995–1007 1941
13 Daschagaran A D Atlas on Congenital Malformations of the Heart Moscow, Dr A A Vishnevsky Clinic 1957
14 Manion W C Personal communication

CHAPTER XXVII

COARCTATION OF THE AORTA

COARCTATION of the aorta means that there is a constriction in the aorta which obstructs the flow of blood through it The constriction may occur anywhere, by far the most usual location is close to the point of entrance of the ductus arteriosus The coarctation may occur above the point of entrance of the ductus arteriosus, that is, it may be *preductal,* or it may occur below the entrance of the ductus arteriosus, that is, it may be *postductal* Occasionally it may occur in the thoracic portion of the aorta and in rare instances a constriction is found in the abdominal aorta

The constriction varies not only in position but also in length The difference in the length is so marked that there are virtually two types of coarctation, the *infantile* and the *adult type,* so named because the former is the type most commonly found in infants dying of the condition whereas the latter is compatible with a long and active life In the *infantile type* there is a diffuse narrowing of the isthmus of the aorta between the left subclavian artery and the point of entrance of the ductus arteriosus In the *adult type* there is a localized constriction which usually occurs close to the point of entrance of the ductus arteriosus In both, the degree of narrowing is subject to marked variation In either, the constriction may be so extreme as to cause complete interruption of the descending aorta In the *infantile type,* if the constriction is so marked as to cause complete interruption, the entire aorta between the left subclavian artery and the point of entrance of the ductus arteriosus is obliterated, the circulation to the lower extremities is from the pulmonary artery through the ductus arteriosus to the descending aorta The resulting condition is no longer a coarctation but a complete interruption of the isthmus of the aorta (see Chapter xii) In the *adult type* the *situation is quite different even if there is complete interruption*, the ductus arteriosus undergoes normal closure and the blood from the ascending aorta reaches the descending aorta by the devious pathways of the collateral circulation

Furthermore, coarctation of the infantile type is frequently associated with some gross malformation of the heart which in itself is not long compatible with life, whereas coarctation of the adult type commonly occurs as an isolated malformation and is readily compatible with life

The physiological difference between the two types of coarctation, as emphasized by Johnson et al,[3] depends on the point *of entrance* of the ductus arteriosus By definition a coarctation of the *infantile type* is always *preductal*, [illegible] may or may not be *postductal* In a pre

[illegible]

lower extremities is from the pulmonary artery [illegible] the other hand, the ductus arteriosus undergoes normal obliteration the only possible pathway by which the circulation to the lower extremities can be established is by the collateral circulation Consequently after the obliteration of the ductus arteriosus there is no difference between the various types of coarctation The signs and symptoms are the same, they result from the constriction of the aorta

ETIOLOGY

The etiology is unknown It is even possible that the etiology of the two types of coarctation may be different. One widely accepted theory of the etiology of coarctation of the aorta of the *adult type* is the Skodaic theory,[*] which postulates that there is an extension of the tissue of the ductus arteriosus into the adjacent portion of the aorta and that, as the ductus arteriosus undergoes normal obliteration, the tissue lying in the wall of the aorta also contracts and thereby causes the coarctation If this theory is true, it means that the constriction of the aorta occurs after birth

If the collateral circulation to the systemic circulation is dependent solely upon extra uterine conditions, there is an extraordinary difference between the ease with which adequate collateral circulation develops in the systemic circulation as compared with that in the pulmonary circulation In cases of pulmonary atresia, even though the obliteration of the ductus arteriosus may be delayed, the bronchial arteries seldom become sufficiently dilated for the maintenance of life Therefore, it seems to the author probable that adequate collateral circulation to both the systemic and the pulmonary circulation develops only if, during intra uterine life, the circulation is such as to initiate the formation of these pathways If the constriction dates from intra uterine life, it means that the ductus arteriosus enters the aorta at or above the point where the aorta is abnormally small Under such circumstances not only is the flow of blood from the ascending aorta to the descending aorta reduced, but also little blood can reach the descending aorta by way of the ductus arteriosus This would cause a great reduction in the circulation to the trunk and the lower extremities If the circulation to the body

were insufficient to meet the needs of the growing embryo, the pathways of collateral circulation would develop during intra uterine life

In the *infantile type* of coarctation of the aorta the situation is quite different By definition the constriction occurs between the left subclavian artery and the point of entrance of the ductus arteriosus Under such circumstances the main circulation to the lower extremities is from the pulmonary artery through the ductus to the descending aorta Usually the ductus arteriosus is of normal size or abnormally large and the circulation to the trunk and the lower extremities is adequate, hence there is no stimulus for the development of collateral circulation

The diffuse narrowing of the isthmus of the aorta suggests that during early embryonic life there was a slight shift in the point of separation of the blood streams to the upper and the lower extremities The alteration in the stress and strain was insufficient to cause a complete interruption of the aortic arch but was sufficient to cause a decrease in the flow of blood through the aorta Therefore the isthmus of the aorta remained hypoplastic This theory of alteration in stress and strain at the time the vascular tree was undergoing differentiation would also explain the frequency with which this malformation is associated with gross abnormalities of the great vessels and the manner in which they arise from the heart

NATURE OF THE MALFORMATION

The essential feature of a coarctation of the aorta of the *adult type* is a localized constriction of the aorta which most commonly occurs at the point of entrance of the ductus arteriosus, as shown in Figure XXVII–1 The constriction may be so extreme that no blood can flow through it to the descending aorta In spite of this, the ductus arteriosus undergoes normal obliteration in some cases a cord like remnant remains, in others only a dimpling in the aorta persists to indicate its former location The circulation to the lower extremities is through the devious pathways of the collateral circulation

Occasionally the constriction of the aorta occurs above the origin of the left subclavian artery, thus rendering the pulse in the left arm weaker than that in the right In rare instances the reverse has been known to occur the left subclavian artery is given off above the constriction and the right subclavian artery arises anomalously from the descending aorta below the level of the constriction, thus the pulse in the left arm is strong and that in the right is weak or absent [3] Indeed, occasionally the right subclavian artery may arise from the left subcla



FIGURE XXVII–1 Coarctation of the aorta of the adult type

vian artery, which in turn arises below the constriction, thus rendering the patient pulseless in all four extremities In rare instances the constriction may occur in the abdominal aorta Regardless of the site of the constriction, the anomaly affects the aorta the heart itself is normally formed with the single exception that the aortic valve may be bicuspid

The basic feature of a coarctation of the aorta of the *infantile type* is a diffuse narrowing of the isthmus of the aorta between the left subclavian artery and the point of entrance of the ductus arteriosus, it is an exaggeration of the narrowing of the aorta which normally occurs in this region Therefore the condition should not be diagnosed as abnormal unless the aorta narrows approximately to the width of one of the carotid arteries Figure XXVII–2 illustrates the usual location of the narrowing of the aorta in the infantile type of coarctation

Although the ductus arteriosus is frequently abnormally large and serves as the main pathway by which blood reaches the trunk and the lower extremities, in some instances—notably when the coarctation occurs as an isolated anomaly—the ductus arteriosus is abnormally small Under such circumstances the pathways of collateral circulation develop during intra uterine life

When coarctation of the aorta of the *infantile type* occurs as an isolated malformation, the small "coarcted" segment may dilate as the ductus arteriosus undergoes obliteration, so that as the individual grows this type of coarctation may ultimately come to resemble a coarctation of the aorta of the *adult type* (compare Figures XXVII–1 and 3)

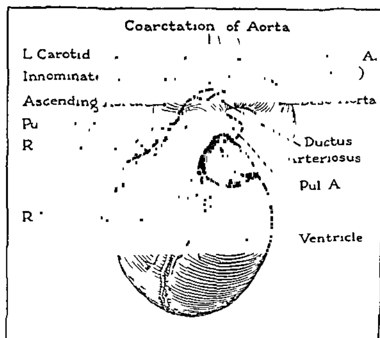


FIGURE XXVII-2 Coarctation of the aorta of the infantile type (preductal coarctation)

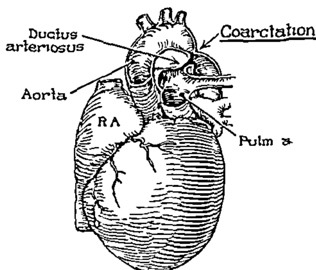


FIGURE XXVII-3 Preductal coarctation of the aorta

Even when the coarcted segment is relatively long, if the ductus arteriosus is of normal size or of small caliber and undergoes normal obliteration, the condition is functionally identical with that of coarctation of the aorta of the adult type Indeed, the case of a fourteen year-old boy reported by Hamilton and Abbott[4] as coarctation of the adult type is anatomically a coarctation of the infantile type in which the ductus arteriosus had closed and there was extensive collateral circulation

The collateral circulation is developed from three principal anastomotic channels The first is around the apex of the thoracic cage, the second is by way of the shoulder girdle, the third is through the internal mammary artery All three of these pathways receive part or all of their blood from branches of the subclavian artery *

The apex of the thoracic cage is of great importance because the aorta arches posteriorly from the level of the second anterior costal cartilage to the level of the fourth thoracic vertebra The consequence is that the apex of the thoracic cage is supplied in part from the ascending aorta and in part from the descending aorta

The two principal pathways by which the collateral circulation is established around the apex of the thoracic cage are first, by way of the superior intercostal artery and second, by way of the inferior thyroid artery

The first thoracic space is supplied solely from the superior intercostal artery, which arises from a branch of the subclavian artery and therefore takes no part in the collateral circulation The second thoracic intercostal space is supplied in part by the superior intercostal artery, which arises from the ascending aorta, and in part by the first aortic intercostal artery, which arises from the descending aorta below the level of the constriction These two arteries anastomose and thus they establish a direct communication between the ascending and descending aortae The second pathway around the apex of the thoracic cage is established

* The subclavian artery normally has three branches of great importance in the collateral circulation These are the thyrocervical trunk the internal mammary artery and the costocervical artery The last mentioned is a short trunk which soon divides into the *deep cervical* artery and the superior intercostal artery The thyrocervical artery is also a short trunk, which almost immediately divides into the inferior thyroid artery, the transverse cervical artery and the transverse scapular artery The trunk of the *thyrocervical artery* is frequently absent under such circumstances the three arteries arise directly from the subclavian artery Thus although subject to slight anatomical variations functionally these vessels are all branches of the subclavian artery

between a branch of the inferior thyroid artery, which receives blood from the subclavian artery, and the posterior branch of the first aortic intercostal artery

The shoulder girdle is probably the most important pathway by which the collateral circulation is established The main circulation to the shoulder girdle is from the transverse cervical artery, the transverse scapular artery, and the subscapular artery The first two are branches of the subclavian artery, the third arises from the axillary artery Thus all three vessels receive their blood from the ascending aorta

The transverse cervical artery arches anteriorly above the clavicles, where it may produce conspicuous pulsations The descending branch of this artery, which lies posteriorly and supplies the vertebral border of the scapula, frequently becomes dilated and causes conspicuous pulsations in the interscapular region The axillary portion of the shoulder girdle receives its blood supply from branches of the subscapular artery and the transverse scapular artery, the latter gives off the circumflex scapular artery and thoracodorsal artery These two arteries emerge from the axilla and have many anastomoses with the transverse scapular artery It is the enlargement of these arterial pathways which causes the pulsations frequently visible below the scapula and in the axilla This rich network of anastomosing arteries gives off branches which pierce the intercostal spaces from behind and anastomose with the second and sometimes with the fourth intercostal arteries, blood is thereby poured from the ascending aorta by way of the subclavian artery and the shoulder girdle into the descending aorta below the level of the constriction (see Figure xxvii-4)

Branches from the axillary artery to the anterior chest wall also help to establish the collateral circulation The thoraco-acromial artery, a branch of the axillary artery, anastomoses with the transverse scapular artery and with the lateral thoracic artery (the long thoracic artery) and also with the lateral branches of the intercostal arteries

The internal mammary artery is the third principal pathway by which the blood from the ascending aorta reaches the lower extremities As the internal mammary artery extends down the abdominal wall, it becomes the superior epigastric artery The superior epigastric artery in turn anastomoses with the inferior epigastric artery, which arises from the external iliac artery By this pathway a direct anastomosis is established from the ascending aorta to the iliac artery and the lower extremities This is also shown in Figure xxvii-4 The internal mammary artery itself arises either from the thyrocervical artery or directly from the subclavian artery and passes anteriorly immediately beneath the

Figure XXVII–4 Collateral circulation in coarctation of the aorta of the adult type

sternum where it gives off branches to the anterior intercostal spaces Since the first and second posterior intercostal spaces also receive their blood supply from the subclavian artery, these anterior intercostal arteries are never dilated The lower anterior intercostal arteries, those to the fourth, fifth, and sixth intercostal spaces, anastomose with the corresponding posterior intercostal arteries, which receive their blood supply solely from the descending aorta, consequently these anterior intercostal arteries may become dilated

Inasmuch as the principal pathways of the collateral circulation are by way

of the posterior intercostal arteries, these arteries become enormously enlarged and tortuous The tortuous loops of the intercostal arteries press against the lower margins of the ribs, as shown in Figure xxvii–5 The pulsation in these loops is so marked that in the course of years these arteries erode the ribs and produce a scalloping of the lower margins of the ribs The erosion of bone is a slow process Although occasionally notching of the ribs may be present by three or four years of age, it is usually not apparent until twelve or fourteen years of age

The collateral circulation increases as the patient grows Over a period of years it becomes progressively more conspicuous For this reason the classic clinical picture may not become clearly manifest until early adult life

COURSE OF THE CIRCULATION

During fetal life the constriction of the aorta alters the course of the circulation in that blood cannot flow freely from the ascending aorta to the descending aorta If the coarctation of the aorta is of the *infantile type* it alters the course of the circulation only insofar as less blood flows from the ascending aorta to the descending aorta and more blood flows from the pulmonary artery through the ductus arteriosus to the descending aorta The alteration in the circulation may cause the ductus arteriosus to enlarge but otherwise the malformation places little strain on the circulation The course of the circulation is shown in Figure xxvii–6

If, however, the ductus arteriosus is of normal size and the constriction of the aorta is great, as shown in Figure xxvii–7, or if the ductus arteriosus opens into a coarcted segment, as is also common in a coarctation of the adult type, difficulty in the direction of blood to the trunk and the lower extremities is encountered and the pathways of collateral circulation enlarge These pathways compensate for the obstruction in the aorta The strain on the fetal circulation is minimal, at birth the heart is normal in size

After birth, if the coarctation is preductal, the expansion of the lungs does not lead to the normal reversal in the direction of the flow of blood through the ductus arteriosus Although more blood flows from the pulmonary artery to the lungs than did during fetal life, the constriction of the aorta means that the pressure in the descending aorta remains low and blood continues to flow through the ductus arteriosus to the descending aorta Thus the blood from the right auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs and through the ductus arteriosus to the trunk and the lower

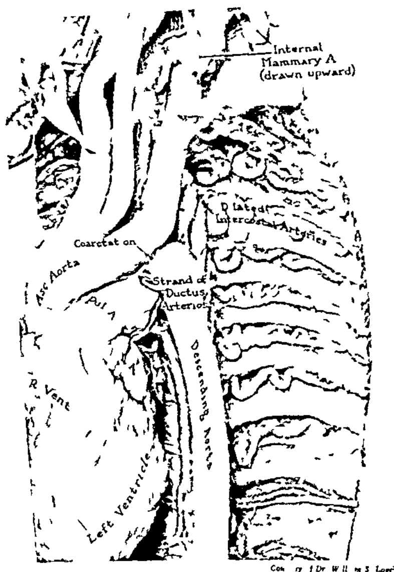


Figure xxvii–5 Coarctation of the aorta of the adult type and the collateral circulation

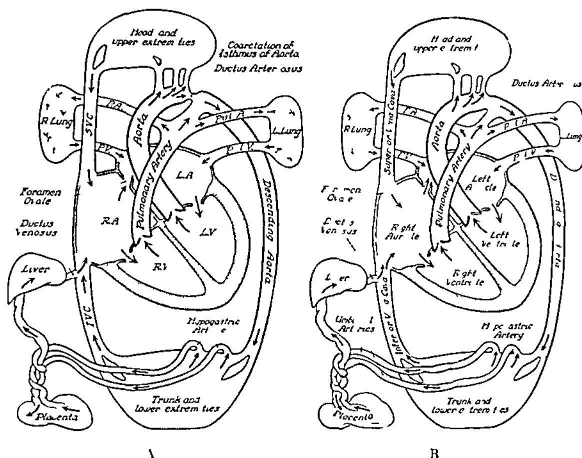
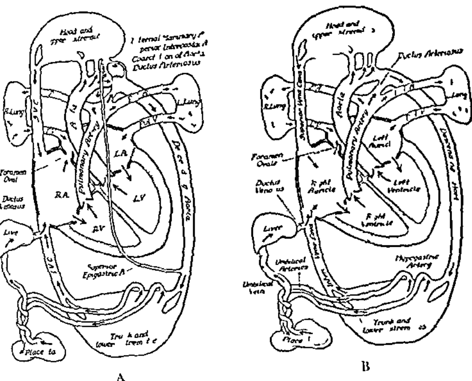


FIGURE xxvii-6 Fetal circulation (A) Coarctation of the aorta of the infantile type and (B) normal heart

extremities The blood which flows to the lungs is oxygenated and returns in the normal manner to the pulmonary veins and the left auricle Thence it flows to the left ventricle and is pumped out into the aorta Most of the blood in the aorta flows to the head and the upper extremities and a small volume of blood flows through the coarcted segment to the descending aorta, where it meets the blood which flows through the ductus arteriosus Thus the upper extremities receive fully oxygenated blood and the lower extremities receive a mixture of oxygenated and venous blood The blood from the body is returned in the normal manner by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again (see Diagram xxvii-1)

When the constriction of the aorta is postductal or, indeed, if the ductus arteriosus undergoes normal obliteration, although difficulty may be encountered in the establishment of the circulation, the only alteration in the circulation is the manner in which the blood reaches the trunk and the lower extremities Thus the blood from the right auricle passes to the right ventricle and is pumped



A B

FIGURE XXVII-7 Fetal circulation (A) Coarctation of the aorta of the adult type and (B) normal heart

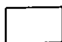
out through the pulmonary artery to the lungs The blood from the lungs is returned to the left auricle and thence to the left ventricle The blood from the left ventricle is pumped out into the aorta Although difficulty is encountered in the flow of blood to the descending aorta, all the blood which flows to the head and the upper extremities is returned by the superior vena cava and all the blood from the trunk and the lower extremities is returned in the normal fashion by the inferior vena cava to the right auricle The only abnormal finding is the manner in which the blood reaches the descending aorta, that is, by the pathways of the collateral circulation The course of the circulation is shown in Diagram XXVII-2

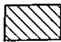
PHYSIOLOGY OF THE MALFORMATION

In early infancy prior to the closure of the ductus the relation of the coarctation to the point of entrance of the ductus arteriosus is of prime importance If the ductus arteriosus opens into the aorta at or above the level of the constriction,

Diagram XXVII-1

Head and upper extremities
Isthmus of Aorta
Ductus Arteriosus
R Lung
P.A.
S.V.C.
Aorta
Pulmonary Artery
Pul. A.
L Lung
PV
Pul V
L A
R.A.
L V
R.V.
I.V.C.
Descending Aorta
Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis


Venous blood

DIAGRAM XXVII-1

Preductal coarctation of the aorta (infantile type)

In a preductal or infantile type of coarctation of the aorta, the ductus arteriosus opens into the aorta below the coarcted segment This means that, as long as the pressure in the lower extremities remains lower than that in the pulmonary artery, blood will flow from the pulmonary artery through the ductus arteriosus to the descending aorta Hence there is pulmonary hypertension

The blood from the right auricle flows into the right ventricle and from there it is pumped into the pulmonary artery Part of the blood in the pulmonary artery flows to the lungs where it is oxygenated, and is returned in the normal manner to the left auricle Thence it flows to the left ventricle and is pumped out through the aorta to the body The blood from the head and the upper extremities is returned by the superior vena cava to the right auricle. The blood to the trunk and the lower extremities passes through the narrowed segment of the aorta to the descending aorta where it is augmented by that part of the blood from the pulmonary artery which flows through the ductus arteriosus to the descending aorta Hence a mixture of arterial and venous blood flows to the trunk and the lower extremities and is returned by the inferior vena cava to the right auricle. There the cycle starts again Thus with each cardiac cycle, the right auricle and the right ventricle receive more blood than was pumped out from the left ventricle during the previous cardiac cycle. Consequently the malformation places an ever increasing load on the right side of the heart

Inasmuch as the head and the upper extremities receive fully oxygenated blood from the left ventricle, they are of normal color the lower extremities receive a mixture of oxygenated blood from the left ventricle and venous blood from the right ventricle, consequently if sufficient venous blood flows to the lower extremities, they will be cyanotic If cyanosis is present the line of demarcation lies at the brim of the pelvis

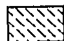
Clinical diagnosis The diagnosis may be difficult. If the baby is seen on the first day of life the pulse in the lower extremities is usually absent By the second or third day of life the circulation to the lower extremities may be sufficiently well established by way of the ductus arteriosus to produce a palpable pulse in the lower extremities Although the lower extremities receive a mixture of venous and arterial blood the amount of reduced hemoglobin in the circulating blood is frequently too small to cause cyanosis The condition is often further complicated by the existence of some additional malformation

Diagram XXVII-2

Head and upper extremities
Internal Mammary Art.
Intercostal Art.
R Lung
S.V.C.
P.A.
Aorta
Pul. A.
L Lung
PV
Pul V
Pulmonary Art.
L A
R.A.
L V
Descending Aorta
R.V.
I.V.C.
Superior Epigastric Art.
Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible


Small admixture of venous blood
No visible cyanosis

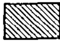

Venous blood

Diagram XXVII-2

Coarctation of the aorta (adult type)

In this malformation there is marked constriction of the aorta distal to the left subclavian artery in the region where the ductus arteriosus enters the aorta. Proximal to the constriction the aorta is usually dilated The narrowing of the aorta is so great that the main flow of blood to the lower extremities is by way of the collateral circulation The main channels of the collateral circulation arise from the branches of the subclavian arteries These are (1) from the subclavian arteries through the intercostal arteries to the descending aorta, (2) by way of the internal mammary arteries through the superior and the inferior epigastric arteries to the femoral arteries

The circulation of the blood is from the right auricle to the right ventricle thence it is pumped out by way of the pulmonary artery to the lungs The oxygenated blood is returned by the pulmonary veins to the left auricle, thence it flows into the left ventricle and is pumped out by way of the aorta to the systemic circulation Inasmuch as the aorta is markedly constricted the blood from the ascending aorta can reach the descending aorta only through the collateral circulation The blood from the upper extremities is returned by the superior vena cava to the right auricle and that from the lower extremities is returned by the inferior vena cava to the right auricle There the cycle starts again

Clinical diagnosis The clinical finding of greatest diagnostic importance is the high blood pressure in the upper extremities combined with a weak or absent pulse in the lower extremities As the patient grows older the collateral circulation becomes more and more marked When the collateral circulation is well developed a systolic murmur is usually audible and pulsations can frequently be detected in the interscapular region The notching of the ribs caused by the increased pressure of the collateral circulation takes time to develop it is usually demonstrable by the time the patient is twelve or fourteen years of age

no great alteration in the circulation occurs, the difficulty is mainly one of adjustment to the increased work required of the left ventricle If, however, the coarctation is *preductal*, that is, the ductus arteriosus opens below the level of the constriction, while the pressure in the descending aorta is abnormally low, blood flows from the pulmonary artery to the descending aorta Under such circumstances, as the author has emphasized, the right auricle receives all the blood which the left ventricle has pumped into the systemic circulation plus that part of the blood from the right ventricle which flows from the pulmonary artery through the ductus arteriosus to the descending aorta Consequently, with each cardiac cycle, the right auricle and the right ventricle receive more blood than was pumped out during the previous cardiac cycle This causes an imbalance between the two circulations, the infant develops right sided cardiac failure The two crucial factors for the restoration of the normal circulation are the rate at which the ductus undergoes obliteration and the time required for the establishment of the normal pressure in the trunk and the lower extremities Although the pulse in the lower extremities remains difficult to obtain, eventually the vasomotor tone of the lower extremities, and indeed the actual pressure in the lower extremities, approaches the pressure in the upper extremities

The establishment of the systemic pressure in the lower extremities may not occur as promptly as it does in the normal individual, but as soon as the pressure in the descending aorta equals that in the pulmonary artery, blood ceases to flow through the ductus arteriosus and a balance is established Nevertheless, as long as the ductus arteriosus remains patent, the pressure in the pulmonary artery is the same as the pressure in the descending aorta, hence there is pulmonary hypertension [5]

After the obliteration of the ductus arteriosus, both the anatomy and the physiology of the various types of coarctation are the same The sole alteration in the hemodynamics is due to the constriction of the aorta which renders it difficult for blood to reach the trunk and the lower extremities and thereby the work of the left ventricle is increased There are no abnormal communications between the two circulations, hence there is no possibility of a shunt

SEX INCIDENCE

The condition occurs far more frequently in males than in females In Abbott's series the ratio of the incidence is four to one,[6] in Lewis' series it is eight to one [7] The reason for this sex difference is not known It is, however, a striking fact that boys with coarctation of the aorta show precocious physical development In contrast, the condition is rare among women of normal physical devel

opment but, as reported by Albright et al[8] and Wilkins and Fleischmann,[9] it occurs frequently among women with ovarian agenesis Wilkins[10] has found that these "women" carry the male sex chromosome Thus it is possible that the etiological factor in this malformation is related to or carried in the male sex chromosome

CLINICAL FINDINGS

There are two distinct clinical syndromes one is seen in infants and the other is found in children and adults It is a striking fact that, although in some instances coarctation of the aorta causes no difficulty in infancy and is frequently not detected until childhood or adult life, in other instances the condition leads to great difficulty in the neonatal period

The clinical syndrome in infants is usually one of respiratory distress which rapidly progresses to severe cardiac failure

Dyspnea may become apparent in the first day or two or may gradually develop during the ensuing week or weeks *Respirations* are rapid and labored There may be *rales* in the lungs

Cyanosis may occur The infant may turn blue with the collapse of circulation and appear as cyanotic as an infant with a venous-arterial shunt, or the lower extremities may appear more cyanotic than the upper extremities

Cyanosis of the lower extremities occurs in the *preductal type* of coarctation because venous blood is pumped through the ductus arteriosus to the descending aorta Under such circumstances the lower extremities will receive a mixture of oxygenated blood from the left ventricle and venous blood from the right ventricle It follows that the lower extremities will be cyanotic, whereas the upper extremities will be of normal color This distribution of cyanosis occurs *in* coarctation of the aorta of the infantile type provided there is no right to-left shunt within the heart When differential cyanosis *is* present, the line of demarcation lies at the brim of the pelvis

The low level of the line of demarcation of cyanosis is due to the fact that the blood supply to the thorax and the abdominal wall is derived mainly from the small branches of the arteries of the shoulder girdle and from the branches of the internal mammary artery and the superficial epigastric artery, all of which are branches of the subclavian artery Therefore the chest and abdominal wall receive their blood supply from vessels which arise above the level of the constriction of the aorta Hence in this malformation these vessels receive oxygenated blood from the left ventricle

This distribution of cyanosis may be both difficult to detect and difficult to

evaluate It is best appreciated by placing the infant's hand beside the foot If, however, there is no cyanosis of the face, lips, hands, and fingernails even after crying, failure to examine the feet with care is a natural omission Moreover, if the feet alone appear to be slightly cyanotic, the usual explanation is that the feet are cold The situation is still further complicated by the fact that this type of coarctation of the aorta is usually associated with some other severe malformation of the heart Frequently the malformation is such that the pressure is higher on the right side of the heart than on the left, under such circumstances any defect in the auricular septum or gross defect in the ventricular septum produces a right to left shunt Such a shunt causes some venous blood to flow into the left ventricle and thus reduces the difference in the oxygen content of the two sides of the heart This lessens or entirely abolishes the difference in cyanosis between the upper and the lower extremities, thereby the differential cyanosis, which is indicative of a coarctation of the aorta above the entrance of the ductus arteriosus, is eliminated

Attacks of abdominal pain may occasionally occur The infant cries out and doubles up his legs Frequently his color becomes ashen gray These attacks are apparently related to the closure of the ductus arteriosus With the spasmodic closure of the ductus the circulation to the abdomen is abruptly reduced Such attacks are probably more common with a coarctation of the adult type or a postductal coarctation before the collateral circulation is well established Be that as it may, difficulty of this nature is quite as serious as is the development of cardiac failure Indeed, the occurrence of such attacks is usually an indication for immediate operation

Cardiac failure which is the most characteristic of all findings, may develop with great rapidity The respirations become labored Râles in the lungs are apparent The liver becomes engorged and edema develops rapidly

The heart is found to be enlarged Usually there is a *gallop rhythm* Occasionally a *presystolic gallop* may be audible over the aortic area A harsh systolic murmur, if present, is usually indicative of an associated malformation

It is the *strong* and *whipping pulse in the upper extremities,* combined with a weak or absent pulse in the lower extremities, which gives the clue to the diagnosis The systolic blood pressure in the upper extremities may be 150 to 200 mm of mercury, whereas in the lower extremities the flush technique gives a blood pressure around 80 to 100 mm of mercury

Infants with cardiac failure should immediately be given digitalis and diuretics If the response is not prompt, or if any evidence of cardiac difficulty per

sists, early operation is indicated (see under Treatment) Hypertension, however, is not an indication for early operation, as it generally subsides during childhood

Fortunately a number of these infants respond promptly to medical treatment The baby rapidly regains compensation The blood pressure gradually falls The heart remains stationary in size as the infant grows Eventually the heart becomes of normal size Thereafter the patient grows and develops as does the patient with a coarctation of the aorta who has shown no difficulty during the neonatal period

Fortunately difficulty in the neonatal period is comparatively rare Most infants are entirely asymptomatic

The clinical findings in children and adults differ markedly from those in infants during childhood these individuals are sturdy and well

The appearance of the patient is often striking Boys and men with this malformation are usually large, strapping, well-developed, sturdy individuals Indeed, the full physical development and the early onset of puberty are quite as distinctive features of this malformation as are the poor physical development and the delayed onset of puberty in patients with an auricular septal defect

Clubbing and *cyanosis* are absent in coarctation of the *adult type* Although difficulty is encountered in the direction of blood to the lower extremities, the blood which does reach the lower extremities is fully oxygenated Inasmuch as there is no abnormal communication between the two circulations, there is no possible chance for an admixture of venous with arterial blood Consequently these patients never show cyanosis or clubbing

The difference in the strength of the pulses in the upper and lower extremities is the outstanding feature of diagnostic importance The strong and powerful pulse in the temporal arteries and in the upper extremities is a striking contrast to the weak or absent pulse in the dorsalis pedis The absence of the pulse in the lower extremities does not mean that blood does not reach the lower extremities but only that it circulates through such circuitous pathways that the force of the heart beat is lost. Lewis[7] has shown that the pulse in the lower extremities is delayed and has a slow rise and a broad plateau

The right radial pulse is frequently stronger than the left This difference in the strength of the pulses in the two arms may occur because the blood reaches the right subclavian artery with greater force Actually in a number of cases autopsy has revealed an anatomical basis for the difference in the blood flow to the two arms In the case reported by Love and Holms[11] there was a marked

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constriction of the left subclavian artery as well as of the aorta Regardless of the cause, whenever there is a difference in the strength of the two radial pulses it is important to palpate the femoral artery

The pulses in all four extremities may, in rare instances, be absent,[3] whereas the pulses in the carotid arteries are strong and vigorous Such an unusual finding should suggest the possibility of a coarctation of the aorta combined with an anomalous origin of the subclavian arteries

The difference in blood pressure between the upper and the lower extremities is of diagnostic significance The blood pressure in the upper extremities is far higher than that in the lower and usually has a *whipping* quality In the lower extremities the blood pressure may be difficult, if not impossible, to record, in many instances no oscillation can be detected on the manometer The systolic blood pressure in the legs is always markedly lower than the systolic pressure in the arms, and the pulse pressure in the legs is narrow The diastolic pressure in the legs, however, is usually higher than the diastolic pressure in the arms The case reported by Railsback and Dock[12] illustrates this phenomenon, the pressure in the arms was 190/90 mm of mercury and that in the legs was 164/140 mm

Hypertension in the upper extremities is common In adults the systolic pressure may be above 200 mm of mercury and the diastolic more than 100 mm The severity of the hypertension is, however, not directly related to the severity of the constriction of the aorta Although some patients with constriction of the aorta suffer from extreme degrees of hypertension, others with similar degrees of coarctation of the aorta show only slight, if any, hypertension

The hypertension in the upper extremities is probably in part caused by the increase in the blood volume in the upper extremities and in part due to the same factors that produce systemic hypertension The decrease in the volume of blood which circulates through the abdomen and the lower extremities may lead to a reduction in the blood supply to the kidneys and to a partial renal ischemia[13, 14] Under such circumstances it seems probable that the slight constriction of the renal artery, which frequently occurs with advancing years, may lead to a far more marked diminution in renal blood flow in patients with coarctation of the aorta than does the corresponding degree of narrowing in normal individuals Similarly, if a patient with a coarctation of the aorta develops essential hypertension, the hypertension tends to become manifest at an earlier age and is more severe than it is in a normal individual

The belief that an underlying tendency to essential hypertension may play

... ion of the aorta is substantiated by the author's obser... quence of events occurs in infants with coarctation of the aorta as in infants with essential hypertension The author has followed a number of patients from birth through adolescence who in infancy showed clinical evidence of essential hyper tension The usual sequence of events is that the hypertension may be extreme during infancy and gradually subside to normal levels during early childhood, and then the patient subsequently develops the so-called hypertension of pu berty Individuals who show such changes in blood pressure in childhood are prone to develop essential hypertension in early adult life The author has stud ied a number of infants with coarctation of the aorta who showed a marked hy pertension in infancy and subsequently had a normal blood pressure during childhood In one instance repeated blood pressure determinations during in fancy were between 200/100 and 180/90 mm of mercury but between five and ten years of age the blood pressure range was approximately 120/80 mm and at twelve years of age the blood pressure had risen to 160/90 mm Regardless of whether these patients again develop hypertension, it is important to realize that an infant with a coarctation of the aorta may show a decrease in hypertension during childhood Furthermore, there is a marked variation in the *severity of* the hypertension which occurs in patients with a coarctation of the aorta It is also important to remember that in childhood and in early adult life the hypertension in the arms is not necessarily extreme Indeed, in some instances the blood pressure in the arms may be normal, namely, 120 systolic and 80 diastolic

Headache and *throbbing* of the head are common in adults and may be very troublesome *Unilateral headaches*,[15] especially if sharply localized, call for careful neurological investigation as they may be indicative of a cerebral *aneurysm*

Plethora and *ready flushing*[7] of the face and neck are relatively common Indeed, when a person suddenly flushes from the *chest upward*, the diagnosis of a coarctation should be suspected Some patients speak of a hot sensation and occasionally of burning of the face and hands Lewis[7] reported one patient who complained that stooping caused a very unpleasant hot feeling in the *head and neck*, so marked that the man avoided stooping under all circumstances'

Numbness and *coldness of the lower extremities are not* uncommon Some patients complain of *weakness in the legs*

Muscular cramps in the lower extremities although *rare*, may occur on severe or prolonged exertion If, however, a patient complains of severe muscle cramps on relatively slight exertion, the possibility that the coarctation involves

the abdominal aorta or that there is a long, diffuse narrowing of the abdominal aorta should always be considered Inasmuch as the collateral circulation is not nearly so easily established in the abdomen as it is in the thorax, patients with this type of coarctation more commonly suffer from poor blood supply to the legs than do those with a thoracic coarctation The exercise tolerance of these persons may be limited by the abrupt occurrence of sharp pain

Difficulty in the healing of wounds in the lower extremities occasionally occurs as a result of the sluggish circulation One of the author's patients complained that, although injuries to the hands and arms healed readily, injuries to the lower extremities always healed slowly The phenomenon was strikingly demonstrated when he suffered a severe burn on his leg which necessitated a skin graft A pinch graft failed to take Indeed, it was not until a skin flap from his abdomen was grafted through his hand, thereby feeding the graft by the circulation of the upper extremities, that the graft on the leg took

Such complaints, however, occur in a minority of cases The vast majority of these patients lead normal, healthy, active lives Not infrequently the condition is first detected upon routine physical examination or by a routine x ray of the chest Indeed, in some instances the condition remains unsuspected throughout life and is found as ' a surprise at the autopsy table

Pulsations can often be detected in unusual places

Pulsations in the episternal notch may be so conspicuous as to suggest an aneurysm

Pulsations in the supraclavicular fossa may be conspicuous and may be seen even in the absence of pulsations in the suprasternal notch Such pulsations are produced by the dilatation of the transverse cervical artery, which arises from the subclavian artery and arches across the shoulder, hence such pulsations may occur in the absence of dilatation of the aortic arch

Pulsations in the vessels in the interscapular region can almost always be felt on careful palpation along the inner margin of the scapula and in the adjacent intercostal spaces Lewis[7] has reported that, if the patient is viewed in a favorable light, these pulsations may be visible posteriorly in the interscapular region, along the margin of the scapula and in the axilla The location of these abnormal pulsations is shown in Figure xxvii–8 The pulsations in the interscapular region are due to the dilatation of the descending branches of the transverse cervical arteries, those at the angle of the scapula and in the axilla are due to the anastomotic pathways arising from the circumflex scapular artery and the thoracodorsal artery, that is, from the subscapular artery and the transverse

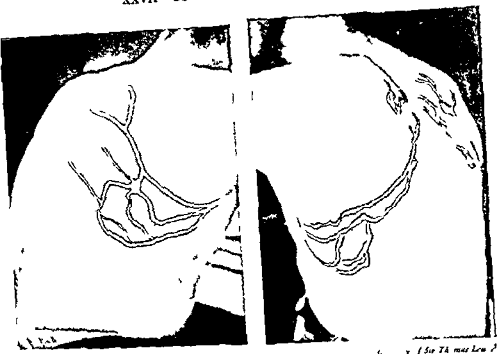


FIGURE XXVII–8 Location of the abnormal pulsations seen in coarctation of the aorta

scapular artery The direction of the flow of blood in all these vessels is into the intercostal arteries and the descending aorta, toward the spinal column King has pointed out that, when these arteries are artificially compressed and emptied of blood, they can be seen to fill from either end, but pulsations occur only when the upper end of the artery is patent

CARDIAC FINDINGS

In early infancy, the *heart* often enlarges rapidly The enlargement is mainly right sided, the condition may progress to severe right sided cardiac failure and may even be fatal

If the infant survives this critical period, compensation is restored and cardiac findings become minimal The heart remains stationary in size as the infant grows, until the normal relation between the size of the heart and the size of the child is established Consequently *in childhood and in adult life* the *heart* is usually *normal* in size Inasmuch as the anomaly concerns the aorta, although the left ventricle may become hypertrophied, there is no cardiac abnormality to produce murmurs or thrills Indeed, murmurs and thrills over the precordium are the exceptions rather than the rule

The aortic second sound may have a ringing, bell like quality, especially if hypertension is severe

A short systolic murmur may occasionally be audible over the aortic area

Both a systolic murmur and a systolic thrill may be present when the aorta is dilated

An early diastolic murmur which is best heard along the left sternal border may become audible as the ascending aorta becomes dilated

A presystolic gallop over the aortic area is common in patients in whom hypertension is severe

Murmurs in unusual places occur in coarctation of the aorta Murmurs may be audible over any of the vessels of the collateral circulation even when these vessels are not sufficiently dilated to be visible Not infrequently, it is the occurrence of a murmur in the interscapular region which suggests the possibility of a coarctation of the aorta In some cases a murmur is audible anteriorly over the internal mammary artery Such a murmur may resemble a precordial systolic murmur but it can usually be heard over both internal mammary arteries, that is, both to the right and to the left of the sternum, and can frequently be traced downward over the abdomen along the course of the superior epigastric artery

Occasionally other dilated vessels of the collateral circulation produce faint thrills and murmurs Abbott and Weiss[6] have emphasized that the occurrence of these murmurs may be due to racemose bunches of dilated capillaries which resemble cirsoid aneurysms They cite cases in which such grape like masses occurred in the axilla, in the brachial artery, and in the abdominal wall

Cardiac failure may occur either in early infancy or in adult life but seldom if ever occurs in childhood

In infants cardiac failure frequently occurs prior to the closure of the ductus arteriosus Indeed, if the ductus opens distal to the constriction of the aorta, there is real danger of right sided cardiac failure because some blood from the right side of the heart which is pumped out into the pulmonary artery flows into the descending aorta and is again returned to the right auricle, thus an ever increasing load is placed on the right side of the heart Fortunately the obliteration of the ductus arteriosus usually occurs sufficiently rapidly to permit the infant to regain compensation Even when the ductus opens proximal to the coarctation, it may cause trouble The author has seen one scrawny infant with evidence of a coarctation of the aorta, a loud continuous murmur, and a wide pulse pressure, who suffered from cardiac failure Subsequently the continuous murmur disappeared, the pulse pressure decreased in width, and the infant regained compensation and thereafter gained weight as a new born infant should The period

of greatest difficulty is prior to and during the period of the closure of the ductus arteriosus

Fortunately many infants who develop cardiac failure and have only an isolated coarctation of the aorta respond to medical treatment and subsequently do well Engle and Goldberg,[16] in their review of a series of infants with coarctation of the aorta, showed that the vast majority of infants who failed to respond to medical treatment had additional cardiac anomalies

In adults the incidence of cardiac failure with coarctation of the aorta is difficult to estimate Hamilton and Abbott,[4] who approached the problem from autopsy studies, stated that 75 per cent of patients with coarctation of the aorta died of cardiac failure before the age of forty King[7] agrees with Hamilton and Abbott. The occurrence of myocardial failure in patients with coarctation of the aorta is, however, not necessarily due to the constriction of the aorta The author cared for a man of forty years of age with a coarctation of the aorta and severe hypertension, a gallop rhythm, and early left ventricular failure Operation corrected the coarctation of the aorta but the hypertension and gallop rhythm persisted Two years later he died suddenly In this instance the patient s difficulty was not primarily due to the coarctation of the aorta but to hypertensive heart disease Lewis, on the basis of his extensive clinical experience, emphasized that coarctation of the aorta, even in the presence of marked hypertension, does not lead to progressive cardiac enlargement and may be compatible with a long and active life

X RAY AND FLUOROSCOPIC FINDINGS

Notching of the ribs is the most characteristic of all features This finding was first pointed out by Railsback and Dock [12] When present, it is readily seen The scalloping, which occurs along the lower margin of the ribs, as shown in Figures XXVII–9 and 10, may be so definite that the diagnosis can be made by this single finding* in the x ray film Erosion of the ribs is a slow process Although it may be apparent by three or four years of age, it is usually not demonstrable until the patient is twelve or fourteen years of age This finding is, however, so constant in older patients with coarctation of the aorta that its absence should always suggest the possibility that the constriction is located in the abdominal aorta (see below)

The heart is usually normal in size The *ascending aorta* may be dilated and

* The only exception to this statement is found in patients who have had a subclavian pulmonary anastomosis After such an anastomosis many patients develop erosion of the ribs on the side on which the subclavian artery has been sacrificed

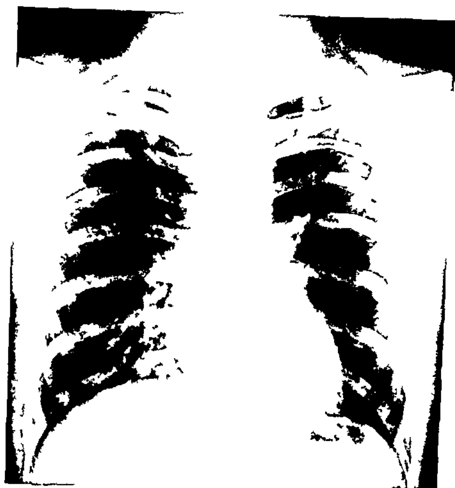


FIGURE XXVII–9 Notching of the ribs in coarctation of the aorta Adult

visible to the right of the sternum The size of the *aortic knob* on the left is subject to great variation In some cases it is strikingly inconspicuous, whereas in others it is unusually prominent (see Figure XXVII–11)

Slight poststenotic dilatation of the descending aorta frequently occurs be low the level of the constriction at the point where the intercostal arteries pour their blood into the descending aorta Occasionally the dilatation of the descend ing aorta is so pronounced that the knob of the aorta is visible at this level, in other instances two aortic knobs may be visible, one lying below the other (see Figure XXVII–12) Such x ray findings, when seen in the anterior posterior film, make the diagnosis of coarctation of the aorta almost as certain as does the oc currence of notching of the ribs

In the left anterior oblique position the ascending aorta is readily visualized Owing to the enlargement of the vessels which arise from the arch of the aorta, the top of the aorta appears to extend abnormally high into the apex of the chest The shadow of the ascending aorta is further exaggerated by the constric



FIGURE XXVII-10 Notching of the ribs in coarctation of the aorta
Enlargement of a portion of the chest shown in *Figure xxvii-9*

tion which renders it difficult to visualize the descending aorta The accentuation of the ascending aorta and the absence of the descending aorta cause the aortic window to be unusually clear (see Figure XXVII-11) Indeed, the contour of the heart in the x ray is not unlike that seen in pulmonary atresia combined with a non functioning right ventricle There is, however, no difficulty in the differentiation of the two conditions The ages of the patients and the physical findings are totally dissimilar

It should be remembered that, when poststenotic dilatation of the descending aorta occurs, it may be best seen in the left anterior-oblique position Under such circumstances, an esophagram shows a slight anterior displacement of the esophagus just below the level of the coarctation

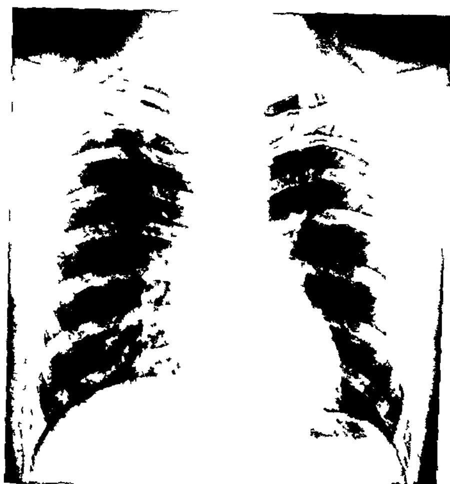


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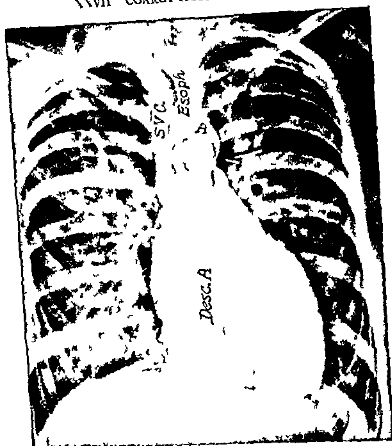
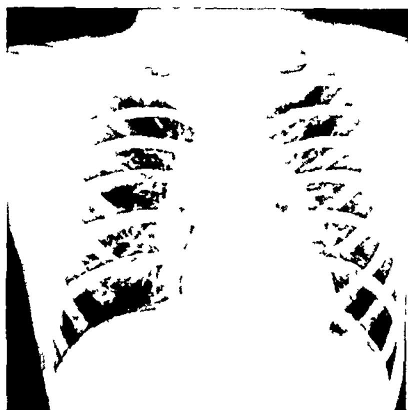


FIGURE XXVII-12. Coarctation of the aorta (x ray) Adult

Arrow points to double aortic knob

ELECTROCARDIOGRAPHIC FINDINGS

In the neonatal period and during early infancy the electrocardiogram usually shows a right axis deviation and evidence of right ventricular hypertrophy During late infancy and early childhood the electrocardiogram becomes balanced and may remain so throughout life In the majority of adults the standard leads show a balanced axis and the unipolar precordial leads show evidence of only a normal degree of left ventricular dominance (see Figure XXVII-13) Evidence of true left ventricular hypertrophy is unusual Indeed, its occurrence is usually an indication for operation Certain it is that evidence of so-called left ventricular strain is rare and that, when it occurs, it is a definite indication for correction of the coarctation



Anterior posterior position



Left anterior-oblique position

FIGURE XXVII–11 Coarctation of the aorta Fifteen years of age

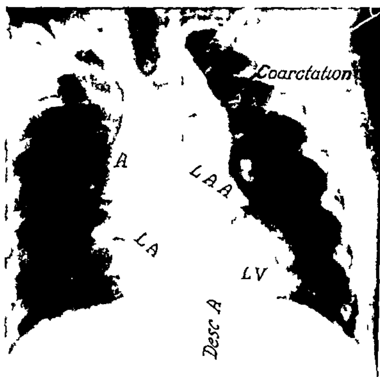




FIGURE XXVII–14 Coarctation of the aorta Adult

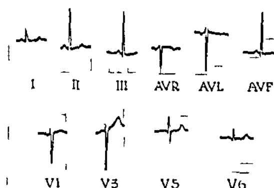


Figure XXVII-13 Coarctation of the aorta Adult

SPECIAL TESTS

Angiocardiography[17 18] usually demonstrates the constriction of the aorta Such demonstration is desirable if there is any question as to the location or the length of the constriction Figure XXVII-14 shows an angiocardiogram of a coarctation of the *adult type*

Retrograde aortography permits the contrast material to reach the coarctation in far higher concentration, consequently this technique gives beautiful visualization of the coarctation (see Figure XXVII-15) The test is, however, not without danger Retrograde aortography in an adult usually requires a relatively high concentration of a contrast substance and the insertion of a catheter through the subclavian artery into the aorta If the catheter is not inserted sufficiently far or if the tip of the catheter slips, there is real danger that upon injection the dye will pass directly up the carotid artery to the brain Dye in high concentration is extremely toxic to the brain and such an accident is almost invariably fatal Compression of the carotid artery at the time of the injection reduces the risk but does not entirely eliminate it If, on the other hand, the catheter is directed down the aorta until it reaches the point of constriction, the aorta may be injured and an aneurysm formed close to the coarcted area Therefore, although this technique offers the best demonstration of the coarctation, the test is not to be lightly undertaken In the author s opinion and in that of Dr Alfred Blalock, this test should be reserved for those rare instances in which there is reason to doubt either the nature, the location, or the length of the constriction Fortunately in most instances far simpler means are available by which the nature of the malformation can be established with certainty

DIAGNOSIS

The diagnosis is based upon a strong pulse in the upper extremities combined

traction on the aorta which is so common in coarctation Indeed, this traction frequently gives the necessary added length which makes an end to-end anastomosis so readily possible

Other causes of cardiac failure in the neonatal period do not cause a difference in the strength of the pulse in the upper and the lower extremities

Essential hypertension is readily differentiated from coarctation of the aorta by the presence of a pulse in the femoral artery Nevertheless, if the femoral artery is not palpated, the coarctation may be overlooked Therefore it is a safe and simple rule never to diagnose essential hypertension without palpation of the femoral artery

Coarctation of the abdominal aorta is a rare anomaly but it does occasionally occur Such a coarctation consists of an abrupt narrowing of the abdominal aorta The narrowed area, which is usually 2 cm or more in length, may include the point of origin of the renal arteries and thereby cause a reduction in the renal blood flow

The patient is usually asymptomatic in childhood, which suggests that the abnormal segment of the aorta does not increase in diameter as the aorta grows, thereby the constriction becomes proportionally greater as the individual attains his growth

As previously mentioned, the possibilities for development of collateral circulation are far poorer in the abdomen than in the chest Consequently the patient is far more likely to suffer from cramp-like pains in the lower extremities than when the constriction is in the usual location (see page 805)

The other signs and symptoms are the same as those of coarctation of the thoracic aorta, with the important exception that notching of the ribs *never* occurs Hence, in an adult who has signs of coarctation of the aorta but *no* notching of the ribs, the possibility of an abdominal coarctation or a diffuse narrowing of the abdominal aorta should be considered Under such circumstances aortography may be indicated

Diffuse narrowing of the abdominal aorta, which usually extends into the femoral arteries, may be confused with a localized coarctation of the abdominal aorta The symptoms and signs are the same as those of an abdominal coarctation, namely, severe cramps in the legs when walking, weak or absent pulsations in the femoral arteries, and *no* notching of the ribs As in the case of coarctation of the abdominal aorta, the condition generally becomes manifest in early adult life, which suggests that this abnormality also prevents the normal growth of the aorta and its main branches

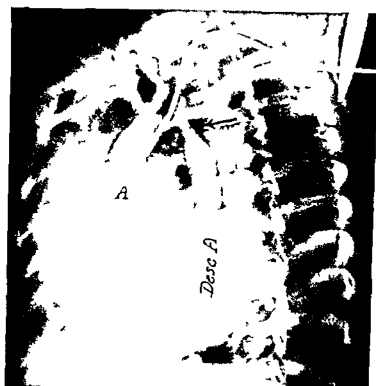


FIGURE XXVII-15 Coarctation of the aorta (aortogram) Infant
Arrow points to the coarctation

with a weak or absent pulse in the lower extremities The diagnosis is confirmed by x ray evidence of notching of the ribs It may also be confirmed by the other evidence of collateral circulation, such as pulsations and murmurs in the inter scapular region

DIFFERENTIAL DIAGNOSIS

In infancy the condition may call for differentiation from a large patent ductus arteriosus and other causes of cardiac failure in the neonatal period In children and adults coarctation of the aorta may require differentiation from other causes of hypertension, from mediastinal tumors, from dilatation of the ascending aorta, and from constriction or narrowing in other portions of the aorta

A large patent ductus arteriosus, in infancy, may simulate a coarctation of the aorta Gross[19] has reported a case in which the ductus arteriosus was so large that it pressed against the aorta and actually constricted the descending aorta to such a degree as to cause obliteration of the pulse in the femoral artery

Patency of the ductus arteriosus occasionally produces a functional coarctation of the aorta [20] The constriction is caused by an exaggeration of the slight

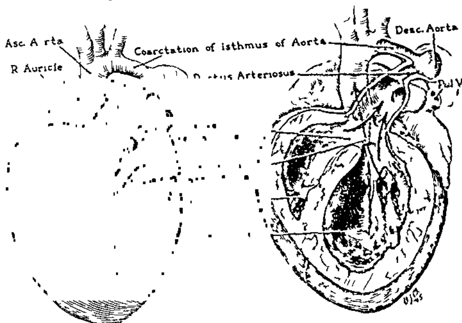


FIGURE XXVII–16 Preductal coarctation of the aorta (infantile type) combined with stenosis of the aortic orifice and over riding of the ventricular septum by the pulmonary artery

gram XXVII–3) Occasionally the left subclavian artery arises from the descending aorta below the entrance of the ductus arteriosus (see Figure XXVII–2) Coarctation of the aorta of the infantile type may also occur in patients with severe left sided cardiac lesions and a reversed ductus (see Chapter XVIII, Section B) When there is a complete transposition of the great vessels with or without a single ventricle, or indeed any malformation which causes persistent cyanosis, and the main circulation to the lower extremities is from the pulmonary artery through a widely patent ductus arteriosus, as shown in Figure XXVII–17, cyanosis is of equal distribution and the pulse in the lower extremities is readily palpable Consequently such an additional anomaly masks the coarctation of the aorta

Endocardial fibro elastosis occurs relatively frequently in infants with coarctation of the aorta Oppenheimer[21] reported a series of such cases in which it appeared that the endocardial fibro-elastosis, not the coarctation, was the probable cause of death Endocardial fibro-elastosis is manifested by evidence of left ventricular hypertrophy and inversion of the T waves in V_5 and V_6 Such a finding is rare in an infant with an uncomplicated coarctation of the aorta When these electrocardiographic findings occur in older individuals, it is difficult to determine whether the changes in the electrocardiogram are caused by the hyperten

Aortography will show the absence of any localized area of constriction, instead there will be diffuse narrowing of the aorta, which usually extends below the bifurcation of the femoral arteries

Surgical correction of this condition is possible, but it requires a long graf which replaces the abdominal aorta, its bifurcation, and the upper portion o both femoral arteries

Mediastinal tumors, especially lymphosarcoma, may compress the aorta and cause a functional coarctation In the presence of a lymphosarcoma or any mediastinal mass, the diagnosis of coarctation of the aorta cannot be made with certainty Under such circumstances treatment should be directed toward the tumor, not to the correction of the coarctation

Aneurysms may be misdiagnosed because of dilatation of the ascending aorta combined with conspicuous pulsations in this region and in the episternal notch Occasionally coarctation of the aorta may be combined with a constriction of one of the subclavian vessels, thereby causing a marked difference in the blood pressure in the two arms This finding is often considered as confirmatory evidence of an aneurysm, consequently the possibility of a coarctation is overlooked and the femoral artery is not palpated The two conditions can be readily differentiated because coarctation of the aorta usually shows the absence of a tracheal tug, the absence of pulsation in the femoral artery, and the presence of collateral circulation

COMMONLY ASSOCIATED MALFORMATIONS

Left sided cardiac lesions occur with sufficient frequency in coarctation of the aorta to suggest that there is more than a casual relation between the abnormalities of the left side of the heart and the abnormality in the wall of the aorta

Bicuspid aortic valves are relatively common in coarctation of the aorta The abnormality causes no difficulty but it does increase the patient s susceptibility to subacute bacterial endocarditis (see Chapter XXXIII, Section A)

Coarctation of the aorta of the infantile type occurs in combination with a wide variety of cardiac malformations *Hypoplasia of the ascending aorta* is common This may occur in combination with a ventricular septal defect, as shown in Figure XXVII–16 Under such circumstances some oxygenated blood is pumped from the left ventricle into the pulmonary artery and more oxygenated blood enters the descending aorta from the ascending aorta, consequently the oxygen saturation in the descending aorta is above the level of visible cyanosis (see Dia

DIAGRAM XXVII-3

Preductal coarctation of the aorta (infantile type) combined with hypoplasia of the ascending aorta and over riding of the ventricular septum by the pulmonary artery

In this preductal or infantile type of coarctation of the aorta the coarctation is combined with hypoplasia of the ascending aorta and an abnormally large pulmonary artery which arises from the right ventricle and slightly over rides the ventricular septum Consequently there is a high ventricular septal defect and some blood from the left ventricle is pumped into the pulmonary artery

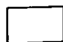
The blood from the right auricle flows into the right ventricle and thence is pumped out through the pulmonary artery to the lungs The blood from the lungs is returned to the left auricle and thence to the left ventricle. Some of the blood from the left ventricle is pumped out through the small aorta to the head and the upper extremities and some oxygenated blood from the left ventricle is pumped out into the pulmonary artery Inasmuch as a large volume of blood is pumped into the pulmonary artery and only a small volume of blood is pumped into the aorta the pressure in the aorta is low Furthermore, because of the coarctation blood cannot flow with ease into the descending aorta, therefore blood from the pulmonary artery continues to flow through the ductus arteriosus into the descending aorta and the pressure in the pulmonary artery is the same as that in the descending aorta Hence there is pulmonary hypertension.

It follows that the head and the upper extremities receive oxygenated blood from the left ventricle and the trunk and the lower extremities receive a combination of oxygenated blood from the aorta and mixed venous and arterial blood from the pulmonary artery For this reason the lower extremities receive an admixture of venous and arterial blood which is at or just above the threshold of visible cyanosis

Clinical diagnosis Clinical diagnosis is extremely difficult as there is no significant difference in the color of the hands and the feet and moreover, the pulse in the lower extremities is readily palpable

DIAGRAM XXVII–3

Head and upper extremities
Coarctation of Aorta
Ductus Arteriosus
R Lung
S.V.C.
P.A.
PV
Aorta
Pulmonary Artery
Pul A
L Lung
Pul V
LA
R.A.
LV
R.V.
Descending Aorta
I.V.C.
Trunk and lower extremities

 Arterial blood (fully saturated)

 Venous and arterial blood
Cyanosis visible

 Small admixture of venous blood
No visible cyanosis

 Venous blood

sternum The murmur is accompanied by a very pronounced systolic thrill Both the murmur and the thrill, whether produced by valvular or subvalvular aortic stenosis, are well transmitted to the vessels of the neck The systolic murmur is also audible posteriorly in the interscapular region and thus may mask a murmur produced by the collateral circulation

Aortic insufficiency causes a loud, ringing, early diastolic murmur over the base of the heart In the author s experience this murmur is quite as well heard along the right sternal border as to the left of the sternum Both the systolic murmur and the diastolic murmur are very pronounced The pulsations in the head and the neck may be conspicuous The radial pulse is of the Corrigan type

Signs of extreme aortic insufficiency and severe aortic stenosis occurring together should always arouse suspicion that the etiology of the valvular lesions is congenital in origin When such signs occur in a patient with a coarctation of the aorta, the valvular difficulty is almost certainly the result of a congenital abnormality

In patients in whom coarctation of the aorta is combined with subvalvular aortic stenosis and aortic insufficiency the contour of the heart is characteristic The heart is enlarged the ascending aorta is dilated, and there is a striking absence of the fullness of the pulmonary conus Although the cause for this change in contour is not obvious, it is a remarkably constant finding The x ray shown in Figure XXVII–18 is an almost exact replica of the x ray of Hamilton and Abbott s proven case [4] In the left anterior-oblique position the dilatation of the ascending aorta is also apparent and the pulmonary window abnormally clear

Rheumatic heart disease must always be considered when there is involvement of the aortic or the mitral valve Palpation of the femoral artery or even of the abdominal aorta reveals the existence of the coarctation This finding does not exclude the coexistence of rheumatic heart disease Either congenital or acquired valvular disease may lead to progressive cardiac enlargement, cardiac failure and death It is the absence of all other evidence of acute rheumatic fever, and indeed the absence of fever and anemia and the persistence of a normal erythrocytic sedimentation rate, which indicates that the valvular abnormality is congenital in origin

COMPLICATIONS

The most serious complications are those directly referable to the high blood pressure in the head and the upper extremities, to weakness of the aortic wall, and to infection

Cerebral vascular accidents may occur They may be caused by the hyperten

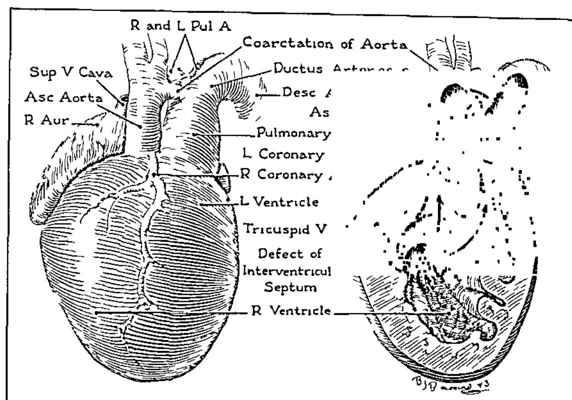


Figure xxvii–17 Preductal coarctation of the aorta (infantile type) with transposition of the great vessels

sion or by the abnormality of the left ventricular endocardium In either case they are serious manifestations

Congenital malformations of the mitral valve produce serious symptoms (see Chapter xxx, Section A) The mitral valve is thickened and may even be displaced downward, as shown in Figure xxx–1 The valve is frequently so malformed that there is insufficiency as well as stenosis The clinical findings are closely similar to those of acquired mitral disease There is a harsh systolic murmur at the apex which is well transmitted to the axilla, in addition, there is usually a readily audible low pitched mid diastolic murmur The malformation places a severe strain on the left side of the heart, the left auricle may become enormously enlarged, as shown in Figure xxx–1, and the left ventricle, too, is greatly dilated and hypertrophied Unfortunately the mitral valve is often so distorted that the condition cannot be corrected by surgery Such a malformation leads to slow progressive cardiac enlargement and eventually to death from cardiac failure

Aortic stenosis and *subvalvular aortic stenosis* (see Chapter xxviii, Section B) both cause a harsh, rasping systolic murmur which is audible over the precordium and of maximal intensity over the base of the heart to the right of the

Aneurysmal dilatation of the descending aorta occasionally occurs in patients with coarctation It is of interest, and probably of significance, that the unusually extreme example of this condition illustrated in Figure XXVII-19 occurred in a young man in whom extensive collateral circulation had developed at an exceptionally early age To the author's certain knowledge it was the presence of a systolic murmur in the interscapular region which first directed attention to the possibility of a coarctation of the aorta when this young man was a child of four A murmur in this location is indicative of the propulsion of blood from the area of high pressure in the ascending aorta through the small collateral vessels into the low pressure area of the descending aorta It is the dissipation of force which causes poststenotic dilatation The development of poststenotic dilatation is a strong indication for surgery In the above instance Dr Frank Spencer corrected the condition by the insertion of a graft

Severe valvular abnormality may seriously increase the load on the myocar



FIGURE XXVII-19 Coarctation of the aorta with poststenotic aneurysmal dilatation of the descending aorta Adult

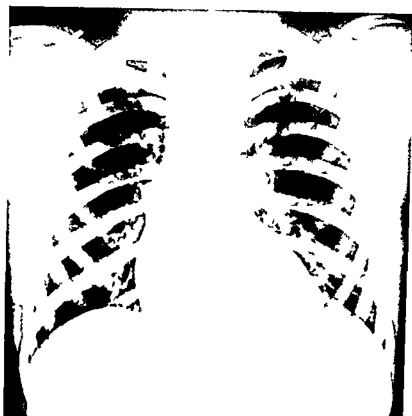


FIGURE XXVII–18 Coarctation of the aorta combined with subvalvular aortic stenosis and aortic insufficiency Child

sion or they may occur because of an associated intracranial aneurysm According to Edwards et al,[22] such aneurysms, which usually involve the circle of Willis, are relatively common in patients with coarctation of the aorta, in that report death was caused by intracranial hemorrhage in approximately 7 per cent of patients with untreated coarctation of the aorta Therefore this possibility should be investigated if a young patient with coarctation of the aorta complains of a unilateral headache or of sharply localized 'migraine headache

Rupture of the aorta is also a danger Usually the rupture occurs proximal to the point of constriction but in a few cases it has been reported to occur below the constriction[23] Because of this risk, strenuous activity, especially that which places a sudden strain on the circulation, is contraindicated

Slight aneurysmal dilatation of the ascending aorta occurs so frequently that it is scarcely considered as a complication

Extreme aneurysmal dilatation of the ascending aorta may develop if a coarctation of the aorta is complicated by medial necrosis (see Chapter XXVIII, Section c) Under such circumstances the high pressure in the ascending aorta may lead to tremendous aneurysmal dilatation of the aorta

other hand, inasmuch as coarctation of the aorta can be corrected by surgery, it is sad to lose a baby with coarctation of the aorta Therefore, if the mortality rate for this operation is low, early operation is often advisable (see below)

Subacute bacterial endocarditis or any suspicion thereof calls for prompt treatment (see Chapter v)

Prophylactic antimicrobial agents should always be recommended prior to and immediately after dental extraction or tonsillectomy Massive chemotherapy or antibiotic treatment should be given whenever a blood stream infection is suspected Antimicrobial substances in high doses are also indicated during the surgical correction of the malformation and should be continued until healing is complete

Even after a coarctation of the aorta has been surgically corrected, prophylactic antimicrobial therapy is still indicated There is always the possibility that the patient may have a bicuspid aortic valve, furthermore, the late occurrence of subacute bacterial endocarditis at the site of the former coarctation, although rare, has been reported[4]

Surgical correction of coarctation of the aorta was developed independently and almost simultaneously by Gross and Hufnagel[5] in Boston and by Crafoord and Nylin[6] in Sweden The operation consists of excision of the coarcted area and end to-end anastomosis of the proximal and distal segments of the aorta, thereby the abnormality is removed Blalock and Park[7] developed an operation to by pass the area of constriction by an anastomosis of the carotid artery or of the left subclavian artery to the descending aorta This operation may be of value if the constriction is so close to the origin of the left subclavian artery that an end to-end anastomosis is not possible and also in cases in which the coarctation is of the infantile type Gross[8] has developed a technique for the insertion of a graft in individuals in whom the area of constriction is too long to permit an end to-end anastomosis

Surgical treatment is indicated in early infancy if the baby does not respond promptly to medical treatment or if he suffers from acute attacks of abdominal pain If a gallop rhythm persists or recurs on slight exertion, or if the infant fails to thrive early operation is indicated As surgical skill increases, early operation for any infant who has suffered from cardiac failure may become advisable It must however, be remembered that the aorta of an infant is a small structure and the sutures probably will not permit the normal growth in the circumference of the aorta at the anastomotic site Therefore there is real danger that the infant will have a significant degree of coarctation when he has grown Further

dium and decrease its nutrition For example, severe valvular or subvalvula aortic stenosis increases the work of the heart and the low systemic blood pres sure decreases the coronary blood flow Indeed, in some instances the calcifica tion of the aortic valve may encroach upon the coronary orifice and lead t coronary insufficiency

Acute and subacute bacterial endocarditis take their toll Inasmuch as th constriction involves the aorta, it is in the systemic circulation that the vegeta tions form Embolic phenomena occur early in the disease The medical treat ment is the same as in other cases of subacute bacterial endocarditis Surgery is contraindicated in the presence of infection, because the suture line of the fresh anastomotic wound is very susceptible to bacterial infection Such infections al most invariably lead to the development of an aneurysm and fatal rupture of the aorta

TREATMENT

Medical treatment is usually preferable for children, because of the danger of abdominal complications prior to the development of adequate collateral circula tion (see below) Virtually the only precaution necessary is to warn the patient against sudden strenuous exertion such as places a sudden excessive strain on the cardiovascular system If the blood pressure is elevated and surgery is to be post poned, this precaution is strongly indicated because of the danger of a cerebral accident or rupture of the aorta Although the majority of these persons lead normal, active lives, the physician will be blamed if the youth drops dead on the athletic field On the other hand, this danger should not be overemphasized A number of men with coarctation of the aorta, in whom the condition was not recognized until later, engaged in active military duty during the first World War with no harmful effects [7] If the patient is asymptomatic, and shows no evi dence either of cardiac strain or of excessive hypertension, no specific treatment is needed

Digitalis and diuretics are always indicated if there is evidence of cardiac failure Even if early operation is indicated, every effort should be made to re store compensation prior to operation Therefore digitalis is always indicated for an infant with cardiac failure Most infants respond promptly to digitalis and diuretics If the infant regains compensation within a day or two and shows no residual indication of cardiac strain, it is advisable not to operate at an early age because of the possibility that the aorta may not grow normally at the site of the anastomosis and consequently the constriction may recur On the

Evidence of slight aortic insufficiency is an indication for operation Such aortic insufficiency is often due to dilatation of the aortic ring Consequently operation not only lessens the load on the heart but may eliminate the dilatation of the aortic ring and render the aortic valve competent

Severe aortic insufficiency and *severe aortic stenosis* are both *contraindications* to operation Unless the aortic insufficiency or the aortic stenosis can be corrected, the relief of the coarctation may lower the blood pressure in the ascending aorta to such an extent as to cause a fatal reduction in the coronary blood flow

COMPLICATIONS FROM OPERATION

The three most common complications are acute bacterial endocarditis, the formation of an aneurysm at the site of operation, and injury to the gastrointestinal tract

Acute bacterial endocarditis may occur after surgery even when the patient receives high concentrations of some antimicrobial substance throughout the early postoperative period, because the fresh anastomotic wound is extremely susceptible to infection There is always the danger of infection from a resistant organism *Pain* in the back in the region of the anastomosis should always arouse suspicion It is quite as significant as the persistence of fever or the development of a hectic temperature When bacterial endocarditis is suspected, a single blood culture should be drawn and immediately thereafter treatment with enormous doses of penicillin or some other antimicrobial agent should be instituted, as

[illegible]

[illegible] may occur if too great tension has been placed on the aorta by the end to-end anastomosis This danger appears to be enhanced by changes in the wall of the aorta which may occur during pregnancy [30] Therefore operation for the correction of a coarctation is contraindicated during pregnancy and pregnancy is contraindicated for the first year after correction of a coarctation

Because of the danger of dilatation of the aorta at the site of the coarctation, it has been a routine practice in Sweden to perform angiocardiography six months after the correction of a coarctation Our experience in Baltimore has shown that an aneurysm of any significant size can be detected by a barium swallow, if the patient is examined in the left anterior-oblique position Therefore we do not believe that a routine angiocardiogram is indicated after operation for a coarctation any more than it is prior to operation

Severe abdominal pain gangrene of the gastrointestinal tract or *perforation*

more, with an infantile type of coarctation, the isthmus of the aorta is so narrow that there is often a significant difference in the size of the isthmus of the aorta and descending aorta (see Figure xxvii–2) Such difference in the diameter of the two portions of the aorta renders a smooth anastomosis virtually impossible, some puckering is almost inevitable Partial relief of the coarctation and ligation of the ductus may, however, lead to a significant decrease in the size of the heart and improvement in the condition of the infant Keith[29] believes that simple ligation of the ductus arteriosus, with special care taken to prevent any distortion of the aorta which might further increase the constriction but with no attempt to resect the coarctation, may be lifesaving for an infant with coarctation of the aorta and cardiac failure, even though the coarctation has to be resected at a later date

If an infant is asymptomatic, he will remain so throughout childhood There fore, if the condition is detected at a routine examination, even though hyperten sion is severe, early operation is not indicated

If possible, it is usually wise to postpone surgery until the child is between ten and twelve years of age, that is, until after the development of collateral circula tion and after the aorta has grown sufficiently to eliminate the danger of serious constriction at the site of the anastomosis and before arteriosclerotic changes in the aorta occur

Shortly after twenty years of age the development of arteriosclerotic plaques may render it difficult to perform an end to end anastomosis For this reason, it is wise to operate before the patient reaches adult life Nevertheless, operation can be safely performed during adult life Indeed, Crafoord, who has operated mainly upon adults over twenty years of age, has had exactly as good results and as low a mortality rate as has Gross, who has operated mainly upon children

Many a patient is so completely asymptomatic that he will question the ad visability of operation

Hypertension or *electrocardiographic evidence of left ventricular hypertro phy or strain* are clear indications for operation in adults

Great cardiac enlargement is usually indicative of an additional anomaly If the anomaly is such as to cause slow, progressive enlargement, as so frequently occurs with anomalies of the mitral valve, the relief of the coarctation removes only one strain It may delay but will not alter the course of events It must also be remembered that the relief of the hypertension will occur only insofar as the hypertension is related to the coarctation Operation will not cure an underlying essential hypertension any more than sympathectomy will cure a coarctation

For patients who show evidence of cardiac strain or hypertension, the prognosis may be greatly improved by surgery

SUMMARY

Coarctation of the aorta means that there is a localized constriction of the aorta This usually occurs in the descending aorta close to the point of entrance of the ductus arteriosus When the constriction occurs above the ductus arteriosus it is spoken of as a *preductal* coarctation in contrast to a *postductal* coarctation in which the ductus opens into the descending aorta above the constriction

The constriction may be sharply localized or there may be a diffuse narrowing of the aorta The former is spoken of as the *adult type* of coarctation When narrowing occurs between the left subclavian artery and the point of entrance of the ductus arteriosus, the condition is known as an *infantile type* of coarctation or a preductal coarctation

Prior to the closure of the ductus arteriosus the relation of the constriction to the point of entrance of the ductus arteriosus has a profound effect upon the hemodynamics of the circulation In a preductal coarctation the blood from the right side of the heart not only flows to the lungs but also flows into the descending aorta and is returned again to the right auricle, this frequently leads to cardiac failure in the neonatal period

Right sided cardiac failure is common in infants with a preductal coarctation If the infant does not respond promptly to medical treatment, surgery is indicated

Attacks of abdominal pain in infancy are also an indication for operation

If the infant survives the closure of the ductus arteriosus, the signs and symptoms are the same, regardless of whether coarctation is long or short

During childhood and early adult life the patient is usually asymptomatic

Patients are sturdy and well developed

Clubbing and cyanosis are absent

The most characteristic of all findings is a strong pulse in the upper extremities combined with a weak or absent pulse in the lower extremities

Occasionally there may be a difference between the two radial pulses and in rare instances the pulses in all four extremities may be absent

High pressure in the upper extremities combined with a low pressure in the lower extremities is characteristic of coarctation

Difficulty in the healing of wounds in the lower extremities may occur

Pulsations may occur in unusual places

The heart is usually normal in size and murmurs are inconspicuous

of the gut may occur in children who have not developed adequate collateral circulation These children complain of severe abdominal cramps, which may be paroxysmal Such patients must be kept under close observation With conservative treatment, the circulation to the intestinal tract may slowly improve and be restored to normal Nevertheless, if the child develops acute intestinal obstruction, signs of perforation, or peritonitis, prompt resection of the injured portion of the intestinal tract is indicated

The author has seen two such complications In the first instance resection of the gastrointestinal tract showed what appeared to be a reaction of periarteritis nodosa, nevertheless, the child made a prompt and complete recovery after the resection of the affected portion of the gut In the second instance the pain gradually subsided and the child spontaneously made an uneventful recovery Two other instances have come to the author's attention one in which operation was performed and no demonstrable lesion found, the other in which operation was postponed too long and the child died from multiple perforations of the gastrointestinal tract Dangers are things to be understood, they are not contraindications to operation

Successful operation restores the circulation to normal The risk of operation is less than 4 per cent In a completely asymptomatic patient, this risk, though slight, may be too great to warrant the effort to restore the circulation to normal

The long time results of operations have been excellent as far as correction of the malformation is concerned Careful follow up studies are needed to determine the long time effect of operation upon the hypertension

PROGNOSIS

The prognosis for an infant with coarctation of the aorta and cardiac failure is guarded Coarctation of the aorta of the *infantile type* carries a high mortality rate in infancy, in part because the condition is frequently associated with other severe malformations of the heart which in themselves are compatible with life for only a short time, in part because the coarctation is preductal and, consequently, until the normal pressure in the lower extremities has been established, the malformation places an ever increasing load on the right side of the heart When there is no additional cardiac abnormality, the prognosis is better

The prognosis for the asymptomatic child or adult with a coarctation of the aorta who has a normal heart, a normal electrocardiogram, and only slight hypertension, is excellent The vast majority of such individuals remain asymptomatic and lead normal, active lives Lewis[7] reported one man with a coarctation of the aorta who lived to the age of ninety two

Most adults desire operation, as the risk is low and operation restores the circulation to normal Severe hypertension, evidence of left ventricular "strain," or evidence of abnormal left ventricular preponderance in the electrocardiogram are clear indications for operation Severe aortic insufficiency and aortic stenosis and even severe mitral stenosis are contraindications to surgery

The most serious complications following surgery are acute bacterial endocarditis, an aneurysm at the site of anastomosis, and gangrene or perforation of the gastrointestinal tract in children

The prognosis for infants with a coarctation of the aorta and cardiac failure is guarded, that for an asymptomatic patient without cardiac enlargement or hypertension is good After operation the prognosis is excellent, as operation restores the circulation to normal

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Cardiac failure may occur in the neonatal period and again in adult life, secondary to long standing hypertension

Notching of the ribs is the most characteristic of the x ray findings It occurs uniformly in older patients In the presence of clinical evidence of a coarctation, its absence should suggest an abdominal coarctation

The electrocardiogram rarely shows evidence of severe left ventricular hypertrophy

Angiocardiography or aortography demonstrate the coarctation

Diagnosis is based upon the difference in the strength of the pulse in the upper and the lower extremities combined with x ray evidence of notching of the ribs

The condition may call for differentiation from other causes of cardiac failure in early infancy, notably patency of the ductus arteriosus In adults coarctation of the aorta may require differentiation from essential hypertension, a diffuse narrowing of the abdominal aorta, or an abdominal coarctation

Left sided cardiac malformations are relatively common in association with coarctation of the aorta Coarctation of the *infantile type* is associated with a wide variety of malformations Coarctation of the *adult type* is often associated with bicuspid aortic valve, endocardial fibro-elastosis, and mitral and aortic valvular abnormalities

The most serious complications are cerebral vascular accidents, severe aneurysmal dilatation of the aorta, rupture of the aorta, and subacute bacterial endocarditis

Medical treatment is always indicated if there is any evidence of infection

Prophylactic antimicrobial agents should always be given prior to dental extraction, even after the coarctation has been corrected, because of the possibility of bicuspid aortic valves or slight abnormality at the site of the former coarctation, either of which renders the patient susceptible to subacute bacterial endocarditis

Digitalis and diuretics are always strongly indicated if there is evidence of cardiac failure

The condition can be corrected by surgery The over all mortality from surgery is less than 4 per cent The ideal time for operation is between the ages of ten and twelve years

The indications for operation in infancy are attacks of severe abdominal pain or cardiac failure which does not respond promptly to medical treatment Hypertension is not an indication for early operation unless it persists into childhood

CHAPTER XXVIII

ANOMALIES OF THE AORTIC VALVE AND OF THE ASCENDING AORTA

ANOMALIES of the aortic orifice may concern only the structure of the aortic valve or they may cause obstruction to the ejection of blood from the left ventricle Occasionally the ascending aorta may be abnormally dilated Section A is concerned with bicuspid aortic valves, Section B with aortic stenosis and Section C with aneurysmal dilatation of the ascending aorta

A Bicuspid Aortic Valves

Bicuspid aortic valves in which the cusps are thin and delicate are clearly congenital in origin Such valves may occur as an isolated abnormality or may occur in association with coarctation of the aorta and other left sided lesions, especially defective development of the left ventricle When the cusps of the valve are thin and delicate, they are usually competent the condition causes no difficulty throughout life

The abnormality is of clinical importance only because of the susceptibility of bicuspid aortic valves to infection[1] Indeed, if subacute bacterial endocarditis occurs in an individual who has no rheumatic history and who shows no clinical evidence of any cardiac abnormality, the possibility of bicuspid aortic valves should be considered

Lewis and Grant emphasized that when bicuspid aortic valves do occur, the two cusps are usually of unequal size and not infrequently a ridge or raphe divides the larger of the two cusps Although the partially divided cusp is the larger of the two it is usually not twice the size of the other cusp

Bicuspid aortic valves and congenitally malformed aortic valves frequently become thickened Calcification of the abnormally thickened valve occurs at an early age and increases still further the obstruction at the *aortic orifice* It is the narrowing of the aortic orifice which causes difficulty, this condition is discussed under aortic stenosis

B Aortic Stenosis

There are a number of different types of abnormalities which interfere with the ejection of blood from the left ventricle The obstruction may occur at the

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CHAPTER XXVIII

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ANOMALIES of the aortic orifice may concern only the structure of the aortic valve or they may cause obstruction to the ejection of blood from the left ventricle. Occasionally the ascending aorta may be abnormally dilated. Section A is concerned with bicuspid aortic valves, Section B with aortic stenosis, and Section C with aneurysmal dilatation of the ascending aorta

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Bicuspid aortic valves and congenitally malformed aortic valves frequently become thickened. Calcification of the abnormally thickened valve occurs at an early age and increases still further the obstruction at the aortic orifice It is the narrowing of the aortic orifice which causes difficulty, this condition is discussed under aortic stenosis

B Aortic Stenosis

There are a number of different types of abnormalities which interfere with the ejection of blood from the left ventricle The obstruction may occur at the

aortic valve or it may occur above or below the valve, that it, it may be valvular, supravalvular, or subvalvular The last mentioned condition is frequently spoken of as *subaortic* stenosis

When the obstruction occurs at the aortic valve, the aortic orifice may be abnormally small or it may be normal in size and the orifice of the valve abnormal The former is usually associated with marked hypoplasia of the ascending aorta and the clinical picture is that of underdevelopment of the left ventricle (see Chapter XIII, Section A) It is the occurrence of aortic obstruction as an isolated malformation which concerns us here

ETIOLOGY

The etiology of valvular aortic stenosis is obscure In adults it may even be difficult to determine whether the obstruction is congenital or acquired In children, however, the congenital types are usually readily differentiated from that caused by rheumatic fever

Subvalvular aortic stenosis is always congenital in origin The stenosis is caused by a band of fibrous tissue or by a muscular ridge which in the adult heart lies approximately 1 cm below the aortic valve

Keith,[3] in his studies of the embryology of the heart, pointed out that a fibrous band is normally present beneath the aortic orifice in early embryonic life and that subsequently it atrophies and disappears He expressed the opinion that subaortic stenosis results from the failure of this membrane to atrophy Furthermore, he pointed out that this is in a sense the counterpart of pulmonary stenosis in the tetralogy of Fallot in which the stenosis is due to the failure of the infundibulum of the right ventricle to expand

In some instances subvalvular stenosis is caused by anomalous insertion of the mitral valve in such a manner as to obstruct the aortic orifice

When subvalvular aortic stenosis is caused by a muscular ridge, the etiology is less clear, but it may be related to the persistence of a remnant of connective tissue band which hinders the normal expansion of the muscular tissue beneath it This is postulated because both may occur together Indeed, in one instance it seemed as if the band of connective tissue had prevented the normal growth and expansion of the left ventricular musculature immediately underlying it and thereby a muscular ridge had been formed which rendered it difficult for the left ventricle to empty itself As the left ventricle hypertrophied, this ridge also hypertrophied and further obstructed the aortic orifice A subvalvular obstruction of this nature is similar to that caused by the muscular hypertrophy which

may occur at the base of the right ventricle in a severe valvular pulmonary stenosis

The etiology of supravalvular aortic stenosis is an enigma

NATURE OF THE MALFORMATION

Supravalvular aortic stenosis is a rare condition which involves the wall of the aorta immediately above the aortic valve When it occurs in an infant or a young child, the wall of the aorta in this area appears puckered and has apparently lost its normal elasticity In an adult there may be a definite ring, as shown in Figure xxviii-1

Congenital valvular aortic stenosis (see Figure xxviii-2) is quite different from congenital pulmonary stenosis Instead of a cap-like valve with a central perforation, only two of the semilunar cusps are fused together and the third raphe remains relatively free Consequently there is a slit like opening This opening uniformly occurs between the non-coronary cusp and the cusp from which the left coronary artery arises and thus causes the aortic valve to appear bicuspid, as shown in insert B of Figure xxviii-2 Nevertheless, in children and young adults the line of fusion between the cusps is usually clearly visible Calcification of this abnormal valve usually occurs in early adult life and may become extreme, as shown in the above mentioned case

Occasionally severe calcification may occur in early childhood The author remembers one case in which the semilunar valves were so completely filled with calcareous material that the base of the aorta appeared to be composed of a solid mass of calcium with only a small central opening Usually, however, the condition is less extreme Nevertheless, it is a condition which tends to increase in severity with age

The increase in severity is in part due to the deposition of calcium in the abnormal valve, which both decreases the flexibility of the valve and further narrows the orifice and also to the hypertrophy of the left ventricle, which causes further hypertrophy of the musculature at the base of the aortic orifice

Subvalvular aortic stenosis or *subaortic stenosis* is caused by the persistence of a band of connective tissue which lies immediately beneath the aortic orifice The obstruction is of this type in approximately 30 per cent of children with signs of aortic stenosis The shelf of connective tissue may stretch across the aortic orifice with only a relatively small opening, as in the case reported by Christian[6] A photograph of this case is shown in Figure xxviii-3 Usually the condition is less extreme The greater part of the membrane atrophies and only

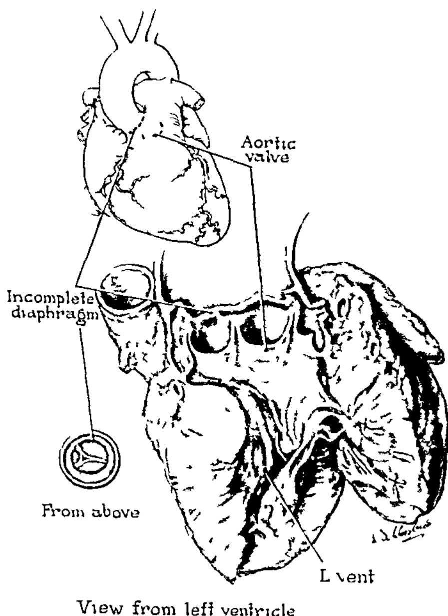


FIGURE XXVIII-1 Supravalvular aortic stenosis Adult

a few bands of connective tissue remain stretched across the base of the aorta, such a condition is known as *partial subvalvular aortic stenosis*

In other instances a subvalvular aortic stenosis is formed by the anomalous insertion of the anterior cusp of the mitral valve beneath the aortic orifice (see Figure XXVIII-4) This is especially prone to occur in association with defects in the membranous portion of the ventricular septum (see also Figure XXII-19)

In many instances the subvalvular obstruction is due to the development of a muscular ridge which narrows the aortic orifice The greater the work required

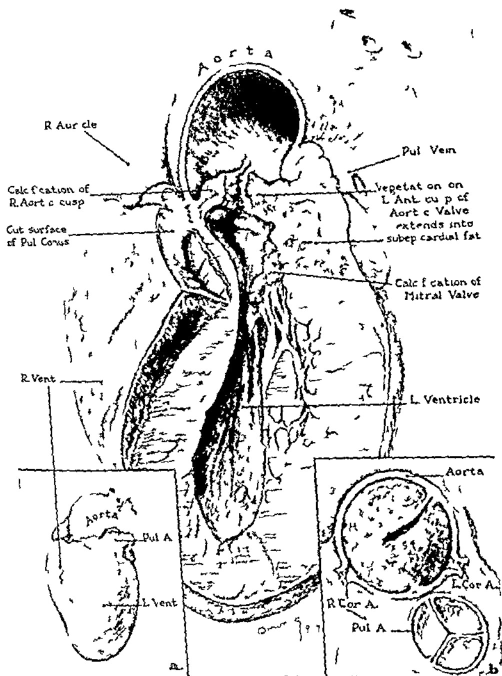


FIGURE XXVIII a Congenital stenosis of the aortic valve Adult

Insert b shows classic fusion of aortic cusps which simulates a bicuspid valve



Courtesy of Dr George Thorn

FIGURE XXVIII–3 Subvalvular aortic stenosis Adult

Note the shelf of connective tissue and the vegetations on the aortic valve

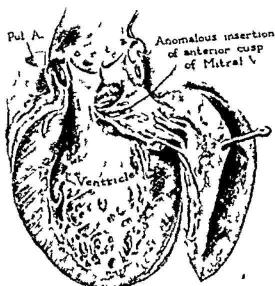


FIGURE XXVIII–4 Partial subvalvular aortic stenosis Infant

Note the anomalous insertion of the anterior cusp of the mitral valve into the aortic shelf

of the left ventricle, the more this muscular ridge hypertrophies and still further obstructs the aortic orifice

Occasionally the combination of a band of connective tissue and a muscular ridge is found The difficulty in the ejection of blood from the left ventricle leads to hypertrophy of the left ventricle and this in turn increases the thickness of the muscular ridge at the base of the aorta Thus a vicious cycle is set up which leads to progressive hypertrophy and progressive stenosis

In rare instances an abnormal muscular ridge lies relatively deep within the left ventricle The etiology of this type of subvalvular obstruction is not known The condition has been termed a functional stenosis, because when the heart is relaxed there is no obstruction, but when the ventricle contracts the muscular ridge obstructs the egress of blood

In all instances the obstruction renders it difficult for the left ventricle to pump the blood into the aorta The heart itself is normally formed but the left ventricle becomes greatly hypertrophied It is, however, worthy of note that, al

though the aortic stenosis leads to great left ventricular hypertrophy, in extreme cases the severity of the aortic stenosis is seldom as great as that which may occur in severe pulmonary stenosis

COURSE OF THE CIRCULATION

The malformation does not alter the course of the circulation The blood from the right auricle passes into the right ventricle and is pumped out by way of the pulmonary artery to the lungs The oxygenated blood from the lungs is returned by the pulmonary veins to the left auricle Thence it flows into the left ventricle and is pumped out through the narrow aortic orifice to the systemic circulation The blood is returned in the normal manner by the superior and inferior venae cavae to the right auricle There the cycle starts again (see Diagram XXVIII–1)

PHYSIOLOGY OF THE MALFORMATION

The obstruction of the aortic orifice increases the pressure against which the left ventricle must work and consequently the pressure in the left ventricle is abnormally high This in turn increases the pressure against which the left auricle must work and the pressure in the left auricle becomes abnormally high The high pressure in the left auricle is transmitted back to the pulmonary capillary bed This affects the circulation in the same way as does severe mitral stenosis The pressure in the right ventricle and that in the pulmonary artery are elevated and the pulmonary wedge pressure is abnormally high The left ventricular pressure increases to compensate for the difficulty in the ejection of blood and consequently for a considerable period of time the systemic blood pressure remains normal As the condition becomes extremely severe, the pressure in the aorta is reduced and consequently the coronary flow becomes less efficient at the time when the myocardium becomes hypertrophied and requires more nutrition These changes further increase the strain on the heart Terminally there is a rapid deterioration in the strength of the left ventricle

SEX INCIDENCE

Congenital valvular aortic stenosis is more common in men than in women The same is true in regard to subvalvular aortic stenosis[6] It so happens that of the three proven cases of supravalvular aortic stenosis with which the author is familiar two occurred in women

CLINICAL FIN

The clin... extreme that the condition is fatal in early infancy or it may ... condition remains inconsequential throughout life Gorlin et al[7] have shown that the condition produces no symptoms until the aortic orifice is reduced to approximately one third of its normal diameter Inasmuch as the initial obstruction is ordinarily less than this, the patient is frequently asymptomatic during childhood Indeed, many patients are asymptomatic until the approach of puberty, when a physiological spurt in growth occurs Furthermore, it is at this age that boys so frequently enter into strenuous sports which greatly increase the work required of the heart. Consequently this is the age at which symptoms frequently develop The development of symptoms is always an indication that the stenosis is severe

Cyanosis is absent, as there is no communication between the two sides of the heart Infants with severe aortic stenosis may show an *ashen color* because of sluggish circulation

Failure to gain weight at the normal rate may also occur in an infant with a severe aortic stenosis Usually, however, the stenosis is not severe, growth and development are normal

Dyspnea is usually absent until the onset of cardiac failure

The pulse pressure varies with the severity of the stenosis Initially the pulse is of normal quality and the *blood pressure* is normal As the stenosis becomes extreme, the blood pressure becomes lower The pulse pressure becomes narrow The pulse is of small volume and has a slow rise and a gradual fall

Fainting attacks and *loss of consciousness* are also late manifestations of severe aortic stenosis The attacks of syncope are due to cerebral anoxia The author was told of one patient who suffered from periodic attacks of loss of consciousness, in this patient autopsy showed that, although the aortic stenosis was not extreme, the valvular deformity was such that the valve could become locked and consequently abruptly cut off the circulation This patient apparently died from an episode in which the valves failed to unlock themselves In other instances syncope is due either to the difficulty encountered in the maintenance of an adequate systemic flow or the patient s inability to adequately increase the cardiac output with exercise

Sudden death is not uncommon Although the aortic stenosis may be extreme autopsy usually does not reveal complete obstruction of the aortic orifice,

DIAGRAM XXVIII-1

Head and upper extremities

R Lung

L Lung

S.V.C.

P.A.

Aorta

Pul A

PV

Pul V

Pulmonary Artery

L A

R.A.

L V

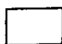
R.V.

Descending Aorta

I.V.C.

Subaortic Shelf

Trunk and lower extremities


Arterial blood (fully saturated)


Venous and arterial blood
Cyanosis visible

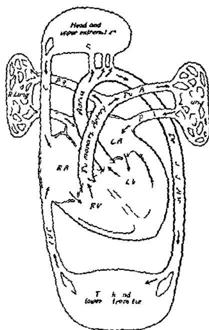

Small admixture of venous blood
No visible cyanosis


Venous blood

DIAGRAM XXVIII-1

Aortic stenosis

The essential feature of th s malformation is an obstruction which may occur at or just below or above the aortic valve. Stenosis of the aortic valve (see insert below) is caused by a fusion of two of the semilunar cusps and thickening and calcification of the valve. Subvalvular aortic stenosis (see diagram on opposite page) is caused by the persistence of a shelf or band of connective tissue which, in the adult, lies about 1 cm below the aortic orifice. Supravalvular aortic stenosis is caused by a narrowing of the aorta just above the aortic valve.



Stenosis of the aortic valve

Regardless of the location of the obstruction, the course of the circulation is essentially normal The blood from the right auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs The blood from the lungs is returned by the pulmonary veins to the left auricle. Thence it flows to the left ventricle and is pumped out into the aorta and the systemic circulation, and is returned in the normal fashion by the superior and inferior venae cavae to the right auricle. There the cycle starts again The stenosis of the aortic valve or the obstruction above or below the aortic orifice increases the work of the left ventricle.

Clinical diagnosis In the early stages that is, when the obstruction is not great the murmur and the thrill closely resemble those of a ventricular septal defect. As the stenosis increases in severity the murmur changes, a har h systolic murmur and a thrill maximal over the aortic area and transmitted to the vessels of the neck develop At this stage the blood pressure i low and the pulse pressure narrow Inasmuch as there is no ab normal communication between the two circulations there is no possibility of a shunt There is no cyanosis and no clubbing

T

cardio

entricu

as in the above mentioned case In many instances the stenosis encroaches upon the coronary orifices Consequently the patient may suffer from coronary insufficiency as well as from cerebral anoxia Indeed, sudden death may be due to ventricular fibrillation

CARDIAC FINDINGS

The heart may or may not be enlarged Inasmuch as the stenotic area grows less rapidly than the normal tissue, the stenosis usually increases with the growth of the individual and thereby increases the strain on the left ventricle The increased work required of the left ventricle to eject the blood through the stenosed aortic orifice leads to hypertrophy of the left ventricle As the left ventricle hypertrophies, the muscular ring at the aortic orifice also undergoes hypertrophy and still further obstructs the aortic orifice Thus a vicious cycle is set up which causes progressive cardiac enlargement As the heart increases in size, although the pressure in the left ventricle rises, the pressure in the aorta falls Consequently, at the time when the musculature of the left ventricle needs increased nutrition, the coronary circulation becomes less efficient

The quality of the second sound at the base of the heart to the right of the sternum may aid in the differentiation of valvular from subvalvular aortic stenosis In the early stage the quality of the aortic second sound is usually normal As the stenosis becomes severe, if it is of the valvular type the second sound over the aortic area becomes weak or absent, whereas if the stenosis is subvalvular in origin, the second aortic sound may be of normal intensity The diminution of the second aortic sound is, however, subject to considerable variation and therefore this finding is not a reliable criterion for the differentiation of the two conditions

A systolic murmur and a thrill are characteristic of this malformation The murmur and often the thrill may be present at birth Initially there is nothing which distinguishes this murmur from that of a ventricular septal defect It is a harsh, rasping murmur which is maximal low down along the left sternal border The obstruction at the aortic orifice sets up a disturbance in the flow of blood which may be extremely widely transmitted, there may be bone transmission of the murmur to the top of the head, to the wrists, and even to the knees At this stage the murmur may be maximal over the aortic area but it is frequently maximal over the precordium in the third and fourth left interspaces The disturbance may be likened to a tumultuous stream flowing rapidly over a rocky bottom While the ventricle is able to pump the blood through the aortic orifice with sufficient force to maintain a normal systemic blood pressure, the disturbance is widely transmitted As the stenosis increases, the murmur becomes

less widely transmitted and more sharply localized over the aortic area Thus, as the patient grows, the character and the location of the murmur and the thrill change[8]

A harsh rough systolic murmur and a thrill, maximal over the aortic area and transmitted to the vessels of the neck, are frequently not found until the child approaches puberty Indeed, these findings, which are so characteristic of aortic stenosis, are usually late manifestations

Aortic insufficiency occasionally develops because of an abnormality of the aortic cusps or dilatation of the aortic ring Its occurrence with subvalvular aortic stenosis is well known but it is not limited to this type of stenosis In the author s experience, when aortic insufficiency occurs in association with congenital anomaly of the aortic orifice, the early decrescendo diastolic murmur characteristic of aortic insufficiency is frequently better heard to the right of the sternum than along the left sternal border

An apical systolic murmur which is transmitted toward the axilla may occasionally be the *only* murmur audible This phenomenon was first reported by Bergeron et al[9] When such a murmur occurs in combination with evidence of severe left ventricular strain, a muscular obstruction deep within the left ventricle should be suspected

Cardiac enlargemen is mainly due to concentric hypertrophy and therefore is not readily apparent for a number of years Nevertheless, cardiac hypertrophy precedes symptoms because, so long as the heart is able to hypertrophy to meet the demands of the body, the patient is asymptomatic Cardiac enlargement usually becomes apparent about the time the *murmur becomes localized* over the aortic area This frequently occurs during the rapid spurt of growth which normally takes place at puberty Unfortunately, as the heart enlarges, *the aortic pressure* becomes lower and the coronary circulation becomes less efficient Consequently there is rapid weakening of the myocardium At this stage a gallop rhythm is common, especially after exertion

Cardiac failure is primarily left ventricular failure Rales in the lungs, pulmonary edema, and cardiac asthma are common The liver also becomes engorged and there may be edema of the extremities Although digitalis will temporarily increase the strength of the heart muscle, eventually the condition leads to intractable cardiac failure and death

X RAY AND FLUOROSCOPIC FINDINGS

The contour of the heart is normal as long as *the heart is normal in size* Inasmuch as the cardiac enlargement is due to left ventricular hypertrophy, as the

heart enlarges the contour becomes boot shaped The pulmonary conus may, however, appear slightly prominent because of the pulmonary hypertension (see Figure xxviii–5) The ascending aorta may appear dilated Such dilatation of the ascending aorta is the counterpart of the poststenotic dilatation of the pulmonary artery which occurs in pulmonary valvular stenosis

Dilatation of the ascending aorta is common in patients with coarctation of the aorta, subvalvular aortic stenosis, and aortic insufficiency In this combination of anomalies in addition to the dilatation of the ascending aorta, there is striking absence of fullness of the pulmonary conus Although the cause for this is not obvious, it is a remarkably constant finding The x ray shown in Figure xxvii–18 is an almost exact replica of the x ray of Hamilton and Abbott's[10] proven case of this combination of anomalies

Examination in the left anterior-oblique position shows that the left ventricle extends abnormally far posteriorly Examination in the right anterior-oblique position may reveal left auricular enlargement

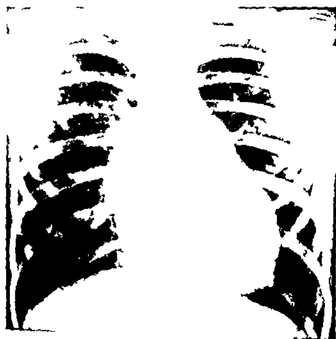
Upon fluoroscopy a hilar dance may be visible, due to the forceful action of the left ventricle, which causes excessive vibration of the heart and great vessels

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiographic findings vary with the severity of the stenosis When the stenosis is not severe the electrocardiogram is frequently within normal limits Evidence of left ventricular dominance in the unipolar precordial leads, however, may occur at an earlier age than is normal Nevertheless, inasmuch as there is considerable variation in the extent of left ventricular dominance found at various ages, no great emphasis can be placed upon this finding

Furthermore, the tendency for the right axis deviation which is so frequently seen in early childhood may persist until the child is ten years of age or older The author has seen one boy who at ten years of age had a tendency to right axis deviation Nevertheless, at this time the unipolar precordial leads showed left ventricular dominance and by fourteen years of age he had evidence of severe left ventricular 'strain"

The electrical axis usually remains balanced until after the left ventricular hypertrophy becomes evident in the unipolar precordial leads Therefore, it is the unipolar precordial leads which offer the clue to the diagnosis When aortic stenosis places a severe strain on the heart, there is inversion of the T waves in Lead I and Lead II and the unipolar precordial leads V_5 and V_6 show a prolongation of the intrinsicoid deflections and tall R waves, combined with depression of the S-T segment and inversion of the T waves, as shown in Figure xxviii–6

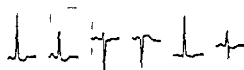


At six years



At twelve years

FIGURE XXVIII–5 Aortic stenosis Child



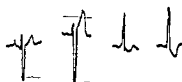


FIGURE XXVIII-6 Subvalvular aortic stenosis Child

SPECIAL TESTS

Cardiac catheterization from the venous side, that is, right heart catheterization *is helpful only* in the exclusion of other malformations, especially a ventricular septal defect

When the left ventricular pressure is greatly elevated, the pressure in the left auricle is also abnormally high and this in turn increases the *resistance in the* pulmonary capillary bed Hence there may be high pressure in the right ventricle and in the pulmonary artery, under such circumstances the pulmonary wedge pressure is always abnormally high

Left heart catheterization is of great aid, as thereby the pressure in the left ventricle can be measured Even if it is not possible to pass the catheter through the aortic valve, the pressure in the femoral artery can be obtained simultaneously with that in the left ventricle and thereby the gradient across the aortic valve can be determined The exact gradient at which operation is indicated cannot be given A systolic gradient of 40 mm of mercury across the aortic valve is usually an indication for operation Nevertheless, it is important to remember that with the advent of left ventricular failure the pressure in the left ventricle may become lower The determination of a gradient across the valve is, however, of great help as occasionally even in the presence of classic signs of aortic stenosis no gradient can be demonstrated The author has seen one such patient, in whom at operation no gradient was found Nevertheless, after operation, at which nothing was known to be done, all evidence of aortic stenosis gradually disappeared and the electrocardiogram returned to normal

Angiocardiography may be of value in the demonstration that, although the left ventricular cavity is not abnormally large, the wall of the left ventricle is abnormally thick It may also aid in the demonstration of the location of the

stenosis (see Figure XXVIII–7) Morrow et al[11] have shown that detailed delineation of the left ventricle and aorta is best obtained by a selective angiocardiogram in which the dye is injected directly into the left ventricle Figure XXVIII–8 shows a supravalvular aortic stenosis in which the origin of the coronary arteries is clearly below the area of stenosis and the ascending aorta appears hypoplastic

DIAGNOSIS

Diagnosis is difficult when the stenosis is not severe, as the systolic murmur is not localized over the aortic area and may be widely transmitted over the body The occurrence of such a murmur combined with electrocardiographic evidence of left ventricular hypertrophy is strongly suggestive of aortic stenosis

A harsh systolic murmur and a thrill over the aortic area which are transmitted to the vessels of the neck are characteristic of aortic stenosis When such a murmur occurs in a patient with cardiac enlargement, a low blood pressure and a narrow pulse pressure combined with electrocardiographic evidence of left ventricular hypertrophy and a pattern of left ventricular "strain," the diagnosis can be made with considerable certainty

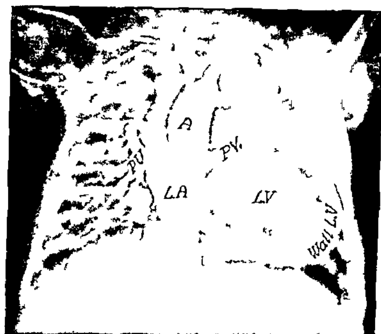
The diagnosis is confirmed by the demonstration that the pressure in the left ventricle is higher than that in the aorta and the femoral artery

DIFFERENTIAL DIAGNOSIS

The condition most commonly requires differentiation from a ventricular septal defect In early infancy the condition also may be confused with a patent ductus arteriosus and occasionally with an endocardial fibro-elastosis, it occasionally requires differentiation from the anomalous origin of the left coronary artery from the pulmonary artery In childhood the condition calls for differentiation from a ventricular septal defect, from an Eisenmenger complex, occasionally from an auricular septal defect and from rheumatic aortic stenosis

A ventricular septal defect requires differentiation from aortic stenosis because in both there may be a harsh precordial systolic murmur and a thrill in a heart of normal size Furthermore, occasionally a large septal defect may be associated with attacks of angina and signs of coronary insufficiency (see Chapter XXIV Section B) The precordial thrill produced by a ventricular septal defect is far more readily palpable than that caused by aortic stenosis Cardiac catheterization readily differentiates the two conditions by the presence or absence of an intracardiac shunt

Endocardial fibro-elastosis is considered because of the electrocardiographic



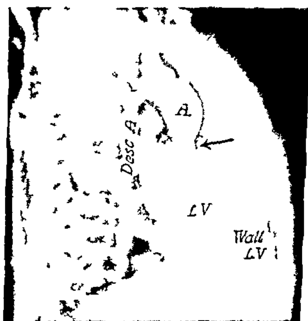


FIGURE XXVIII–7 Aortic stenosis with a thick walled left ventricle Infant

Arrow points to aortic stenosis



Cou sy J D A G Morrow Nat I H a I

FIGURE XXVIII–8 Supravalvular aortic stenosis

evidence of left ventricular hypertrophy, the S-T depression, and the inversion of the T waves in V_5 and V_6 The absence of a significant systolic murmur in endocardial fibro-elastosis differentiates it from aortic stenosis

Patency of the ductus arteriosus in infancy may be associated with a harsh systolic murmur and electrocardiographic evidence of left axis deviation and left ventricular hypertrophy Aortography will reveal simultaneous filling of the aorta and the pulmonary artery If a large ductus arteriosus is demonstrated, the ductus should be closed and subsequently the patient should be re-evaluated The author has known of one infant in whom, upon the ligation of a large ductus, all evidence of a harsh systolic murmur and a thrill over the aortic area disap-

m the pulmonary artery is s usually associated with a harsh systolic murmur The electrocardiograms are totally different in that aortic stenosis shows depression of the S T segment, whereas when the left cor

onary artery arises anomalously from the pulmonary artery there is an elevation of the S-T segment In doubtful cases angiocardiography will clearly differentiate the two conditions, as the left ventricle becomes enormously dilated and its wall extremely thin when the left coronary artery arises anomalously from the pulmonary artery, whereas in aortic stenosis the left ventricular cavity is small and the left ventricle is thick-walled

An Eisenmenger complex with no 'visible" cyanosis may occasionally be confused with aortic stenosis The systolic murmur may be as harsh and as rasping and the thrill as pronounced as in aortic stenosis and in both the murmur is widely transmitted over the entire precordium In the Eisenmenger complex the pulmonic second sound is usually louder than the aortic second sound Moreover, there is no narrowing of the aortic orifice, hence the pulse pressure is normal When an Eisenmenger complex is complicated by aortic insufficiency, the condition may be confused with aortic stenosis combined with aortic insufficiency The sequence of events may be of diagnostic significance If such patients are observed from early childhood, it will be found that the signs of a congenital aortic stenosis precede those of aortic insufficiency, whereas in the Eisenmenger complex aortic insufficiency may be present from early infancy or it may develop insidiously The two conditions are readily differentiated by cardiac catheterization, as there is no shunt in aortic stenosis

An auricular septal defect may occasionally be confused with aortic stenosis when there is the electrocardiographic finding of a right axis deviation and fluoroscopic evidence suggestive of a hilar dance The unipolar precordial leads should differentiate the two conditions, as evidence of a right bundle branch block is characteristic of an auricular septal defect Cardiac catheterization also readily differentiates the two conditions

Rheumatic valvular disease, especially *rheumatic aortic stenosis* calls for differentiation from congenital aortic stenosis The insidious development of rheumatic aortic stenosis in childhood is virtually unknown Usually there is evidence of involvement of the mitral valve, as well as of the aortic valve, and a history of acute rheumatic fever can be obtained Aortic stenosis, rheumatic in origin, is almost invariably preceded by aortic insufficiency Indeed, only once has the author seen a child with such a severe rheumatic aortic stenosis that the signs of aortic insufficiency had disappeared In this patient evidence of mitral stenosis could be elicited Actually the differentiation of rheumatic aortic stenosis from congenital aortic stenosis is more important as regards prognosis than as regards treatment

COMMONLY ASSOCIATED MALFORMATIONS

Dilatation of the ascending aorta and *aortic insufficiency* so frequently occur with a subvalvular aortic stenosis that they are almost part and parcel of the malformation

Coarctation of the aorta is relatively common in combination with aortic stenosis The coarctation of the aorta should never be corrected without the relief of the aortic stenosis (see Chapter xxvii) because the high pressure in the ascending aorta improves the coronary blood flow Indeed, it is usually advisable first to relieve the aortic stenosis and then, at a later date, to correct the coarctation

Severe left-sided cardiac lesion may be associated with aortic stenosis or atresia of the aortic orifice (see Chapter xiii, Section A)

The differentiation between valvular and subvalvular aortic stenosis may be difficult, if not impossible The presence of a normal second sound over the aortic area suggests that the obstruction may be subvalvular Nevertheless, the aortic second sound may be normal even in the presence of a valvular stenosis Furthermore, both conditions may be confused with supravalvular aortic stenosis Angiocardiography is probably the best means to determine the site of the obstruction

TREATMENT

Limitation of exercise is frequently indicated in aortic stenosis, as the condition is one of the few in which cardiac hypertrophy precedes symptoms If it is desirable for any reason to postpone operation in a patient with aortic stenosis and evidence of left ventricular 'strain,' the child s exercise should be markedly restricted Often a period of restricted activity is desirable prior to operation in order to impress upon the child and the parents the severity of the condition

If the diagnosis of aortic stenosis is made prior to the development of left ventricular strain it is wise to limit the child s activity Certainly the child should not be permitted to participate in competitive games or in any activity which places a strain on a normal heart

After the development of symptoms or of electrocardiographic evidence of left ventricular strain exercise should be drastically restricted if for any reason operation is not advisable or feasible

Operation for the relief of aortic stenosis can be done with remarkable success with the use of an extracorporeal circulation [2, 17] This technique permits the surgeon to open the aorta and relieve an aortic stenosis from above It is usually

possible to treat a subvalvular aortic stenosis by a similar approach The results are extraordinarily satisfactory The risk is definitely less than in any operation which requires incision of the left ventricle

Unless there is evidence of complete regression of signs and symptoms and the electrocardiogram returns to normal, moderate limitation of exercise remains a wise precaution even after successful surgery

PROGNOSIS

The prognosis varies with the severity of the aortic obstruction A mild aortic stenosis may be compatible with life for many years One patient with the supravalvular aortic stenosis (see Figure xxviii–1) died in her eighties from un related causes

Usually, however, the prognosis is guarded, as the aortic obstruction increases with age and leads to progressive cardiac enlargement and cardiac failure Un der such circumstances the prognosis is hopeless unless the condition can be re lieved by surgery Successful surgery greatly improves the prognosis

SUMMARY

Aortic stenosis interferes with the ejection of blood from the left ventricle The difficulty may be supravalvular, valvular, or subvalvular Supravalvular aortic stenosis is a rare but well recognized condition Valvular aortic stenosis is caused by the fusion of the right coronary cusp with the two adjacent cusps The entire valve is thickened and subject to early calcification The subvalvular aortic stenosis may be caused by a shelf of connective tissue, by a muscular ring at the base of the aortic orifice, or occasionally by a muscular ridge deep within the left ventricle Subvalvular aortic stenosis occurs in approximately 30 per cent of the cases of aortic stenosis

Any obstruction to the outflow of blood from the left ventricle greatly in creases the work of that chamber The left ventricle undergoes progressive hy pertrophy The course of the circulation is normal The increased pressure in the left ventricle is, however, transmitted back to the left auricle and affects the physiology of the circulation in the same way as does mitral stenosis In addition, as the aortic stenosis becomes extreme, the pressure in the aorta becomes low and interferes with the coronary blood flow

The malformation is more common in men than in women

The clinical findings vary with the severity of the malformation Cyanosis is absent Unless the stenosis is extremely severe, growth and development are nor

mal The clinical symptoms are late manifestations The pulse pressure is narrow The blood pressure is low Fainting attacks and loss of consciousness are serious manifestations Sudden death is not uncommon

The heart may or may not be enlarged The second sound over the aortic area may be diminished A harsh systolic murmur maximal over the precordium, which is widely transmitted, is characteristic of this malformation As the obstruction increases in severity, the murmur becomes less intense, is better localized over the aortic area, and is transmitted to the vessels of the neck An early diastolic murmur characteristic of aortic insufficiency may develop, it occurs more frequently in subvalvular aortic stenosis than in valvular aortic stenosis Cardiac enlargement is due to concentric hypertrophy and therefore it is not easy to detect until it becomes extreme The condition leads to progressive cardiac enlargement and cardiac failure

X ray and fluoroscopy show whether or not the heart is enlarged and whether the aorta is dilated Examination in the left anterior-oblique position gives an indication of the extent of the left ventricular enlargement

The electrocardiogram shows progressive evidence of left ventricular hypertrophy and left ventricular "strain"

Cardiac catheterization shows the absence of a shunt and may show elevation of the pressure in the right ventricle and in the pulmonary artery, and also an elevation of the pulmonary wedge pressure Left heart catheterization gives the pressure in the left ventricle and reveals the extent of the gradient across the aortic valve

Angiocardiography shows the thickening of the left ventricular wall and also aids in the demonstration of the location of the constriction

The condition is to be suspected in a healthy patient with a heart of normal size, a harsh systolic murmur and a thrill, and electrocardiographic evidence of left ventricular hypertrophy The diagnosis is substantiated by the absence of a shunt and proven by the demonstration of a significant gradient across the aortic valve

The condition requires differentiation from a ventricular septal defect In infancy the condition may also be confused with patency of the ductus arteriosus, with endocardial fibro-elastosis, and occasionally with the anomalous origin of the left coronary artery from the pulmonary artery In childhood the condition is frequently confused with a small ventricular septal defect and may be confused with an Eisenmenger complex or even with an auricular septal defect Congenital aortic stenosis requires differentiation from rheumatic aortic stenosis

The commonly associated malformations are dilatation of the ascending aorta with aortic insufficiency, coarctation of the aorta, and severe left sided cardiac lesions

Limitation of exercise is usually advisable Since the condition leads to progressive cardiac enlargement, surgical relief of the obstruction is usually necessary

The prognosis varies with the severity of the stenosis It is usually guarded but can be greatly improved by surgery

C *Aneurysmal Dilatation of the Ascending Aorta*

Aneurysms of the ascending aorta are usually the result of acquired disease, nevertheless, an aneurysmal dilatation of the ascending aorta may be due to a congenital weakness in the aortic wall Such is the nature of the aneurysmal dilatation of the ascending aorta which is characteristic of Marfan's syndrome Since the first case reported by Baer, Oppenheimer, and the author,[18] a number of similar cases have been reported in Marfan's syndrome Indeed, the aortic lesions, together with the skeletal abnormality and the ocular lesions, constitute a veritable syndrome [19] The condition usually appears in more than one individual *in the same family and also in successive generations Congenital aneurysmal* dilatation of the ascending aorta may, however, occasionally occur without any of the stigmata of Marfan's syndrome

NATURE OF THE MALFORMATION

The underlying structural abnormality is primarily due to a deficiency in the elastic tissue of the aorta The lack of a normal amount of elastic tissue causes a weakness of the wall of the ascending aorta, consequently the stretching and thinning of the wall of the aorta lead to medial necrosis An aneurysmal dilatation of the aorta may result therefrom Occasionally a dissecting aneurysm of the aorta occurs Thus, although the aneurysmal dilatation of the aorta is not a congenital abnormality, it is caused by the abnormality in the media of the aorta which is congenital in origin, furthermore, it may be familial and even possibly inheritable

COURSE OF THE CIRCULATION

The dilatation of the ascending aorta in no way alters the course of the circulation

PHYSIOLOGY OF THE MALFORMATION

The physiology of the circulation is normal

CLINICAL FINDINGS

The condition in itself usually causes no symptoms

The appearance of a patient with Marfan's syndrome is, however, characteristic The patient has a tall, slender build and long, tapering extremities (see Figure XXVIII-9), trigger toes are also common Kyphoscoliosis may occur in older individuals and become extreme Ocular disturbances are common, there may be dislocation of the lenses and occasionally detachment of the retina

The occurrence of Marfan's syndrome in several members of the patient's family or in successive generations of his family is strong presumptive evidence that the aneurysmal dilatation of the aorta is due to medial necrosis



FIGURE XXVIII-9 Arachnodactyly in Marfan's syndrome (same patient as in Figure XXVIII-11)

Conspicuous pulsations in the episternal notch may be visible when the dilatation of the aorta involves the transverse arch

CARDIAC FINDINGS

The heart may or may not be enlarged Occasionally there is an increased area of dullness to the right of the sternum due to the tremendous dilatation of the ascending aorta (see Figure xxviii–10)

Displacement of the heart frequently occurs in patients with severe kyphoscoliosis (Figure xxviii–11)

A systolic murmur is common It may be hemic in origin owing to the great dilatation of the ascending aorta or it may be related to displacement of the heart The systolic murmur is frequently so loud as to suggest an additional malformation of the heart

An early diastolic murmur may be audible along the left sternal border, as



Figure xxviii–10 Dilatation of the ascending aorta Adult

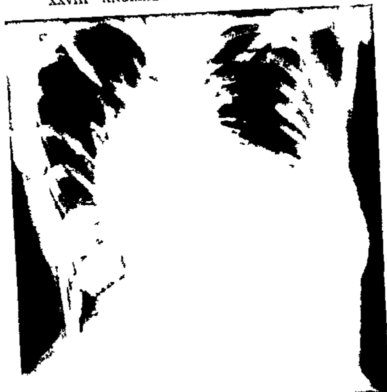


FIGURE XXVIII-11 Congenital aneurysmal dilatation of the ascending aorta in patient with kyphoscoliosis (same patient as in Figure XXVIII-9) Adult

aortic insufficiency may occur as a late complication owing to dilatation of the aortic ring

X RAY AND FLUOROSCOPIC FINDINGS

The condition is usually demonstrated by x ray or fluoroscopy There is dilatation of the ascending arch of the aorta to the right of the sternum The enlargement of the aorta is usually evident in both the anterior posterior and the left anterior-oblique position

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiographic findings depend upon the condition of the heart and are in no way related to the aneurysm of the aorta

SPECIAL TESTS

Cardiac catheterization is of no use in the diagnosis of the abnormality It is, however, of value in the exclusion of any associated intracardiac shunt

Angiocardiography clearly delineates the aorta and thus proves that the shadow is due to dilatation of the aorta and not to a mediastinal mass (see Figure XXVIII-12)

DIAGNOSIS

The diagnosis is usually made by x ray or fluoroscopy It can be confirmed by angiocardiography

DIFFERENTIAL DIAGNOSIS

The condition requires differentiation from aneurysms secondary to acquired disease, from mediastinal tumors, and sometimes from intracardiac malformations

A dissecting aneurysm caused by arteriosclerosis or infection should be excluded before the diagnosis of a congenital weakness of the aortic wall is justified The age of the patient is significant Knowledge that the condition dates from infancy or early childhood or is associated with Marfan s syndrome is strong presumptive evidence that the condition is congenital

Mediastinal tumors may be excluded by the demonstration of expansile pulsations along the margin of the aneurysm The presence or absence of other signs or symptoms of tumors is highly significant Angiocardiography clearly reveals the nature of the condition

Additional intracardiac malformations may be excluded by the normality of the findings on cardiac catheterization

COMMONLY ASSOCIATED MALFORMATIONS

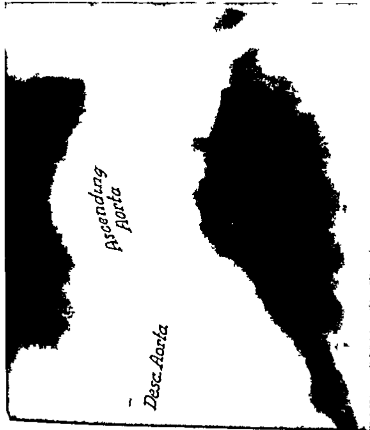
Marfan s syndrome, as previously mentioned, is so common as to complete the clinical syndrome

Coarctation of the aorta may occur When it does occur, the increased pressure in the ascending aorta may accelerate the stretching and dilatation of its wall Therefore, early correction of the coarctation is usually indicated

TREATMENT

Medical treatment is mainly directed toward the prevention of undue strain on the ascending aorta For this reason strenuous physical exercise and that which places a sudden strain on the heart or circulation should be avoided

Surgical treatment is steadily improving and may be of great value It may be advantageous to wrap the aorta with some prosthetic material to prevent



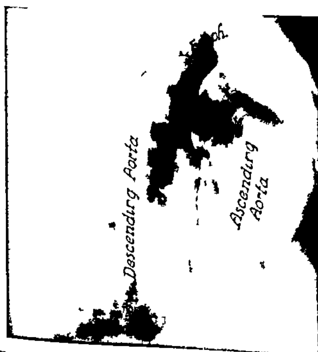


FIGURE XXVIII-12 Congenital aneurysmal dilatation of the ascending aorta Adult

progressive dilatation Dr Frank Spencer has recently inserted a graft to replace the part of the aorta involved in the medial necrosis [20] Such an operation may prove to be of great value

PROGNOSIS

The prognosis, even without operation, is often relatively good Most persons with this condition live until middle life If there is evidence of increasing dilatation of the aneurysmal area to such a degree that there is danger of rupture, successful surgery may eliminate that danger

SUMMARY

Aneurysmal dilatation of the ascending aorta may be the result of a structural weakness of the aortic wall caused by a deficiency of the elastic tissue, this in turn leads to medial necrosis

Medial necrosis of the ascending aorta is common in Marfan s syndrome

The condition usually causes no symptoms but may cause a systolic murmur of sufficient intensity to suggest the possibility of a congenital malformation of the heart This is especially common in patients with Marfan's syndrome and with extreme kyphoscoliosis

The diagnosis is usually made by x ray or fluoroscopy and is confirmed by angiocardiography Cardiac catheterization may be necessary to exclude the existence of additional cardiac abnormalities

The condition, if extreme, may be ameliorated by surgery Even without operation the prognosis is relatively good Most patients with this abnormality live to middle age and in many instances the condition does not shorten the patient s life span

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CHAPTER XXIX

ANOMALIES OF THE CORONARY ARTERIES AND OTHER CONDITIONS WHICH AFFECT THE MYOCARDIUM OF THE LEFT VENTRICLE

THERE are two distinct types of anomalies of the coronary arteries One is caused by the anomalous origin of one or both of the coronary arteries from the pulmonary artery and the other concerns coronary arteriovenous fistulae The former is discussed in Section A, the latter in Section B Subendocardial fibro-elastosis and glycogen storage disease also interfere with the strength of the myocardium, these conditions are discussed in Sections C and D

A *Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery*

One or both of the coronary arteries occasionally arise from the base of the pulmonary artery instead of from the aorta

In 1886 St John Brooks[1] reported two cases in which the right coronary artery arose from the pulmonary artery He made the observation that the two arteries arose from different circulations and therefore received blood under different pressures Consequently he postulated that, as anastomoses develop between the two coronaries, blood must flow in retrograde direction in the coronary artery which arose anomalously from the pulmonary artery In 1911 Abrikossoff[2] published the first report of a case in which the left coronary artery arose from the pulmonary artery Twenty two years later Bland et al[3] suggested that this anomaly produced such a distinctive clinical syndrome that the condition could be diagnosed during the infant s life Since then an increasing number of cases have been reported Soloff,[4] in his review of the literature in 1942, reported seventeen cases in which the left coronary artery arose from the pulmonary artery, whereas, in the remaining four cases, in two instances the right coronary artery arose from the pulmonary artery and in the other two instances both coronary arteries arose anomalously from the pulmonary artery The difference in the reported instances of these anomalies is probably due to the difference in their clinical significance

The anomalous origin of both coronary arteries from the pulmonary artery, according to Soloff,[4] is a rare anomaly and is apparently usually associated with

a malformation of the heart which is so severe that it is seldom compatible with life. A number of cases have been reported in which the right coronary artery arose anomalously from the pulmonary artery, these individuals lived long lives, that is, to seventy four, eighty three, and ninety years respectively, and died from unrelated causes In contrast to this, in the majority of cases of anomalous origin of the left coronary artery from the pulmonary artery, the infants died of cardiac failure between three and five months of age

In former days it was thought that venous blood was adequate for the nutrition of the right ventricle but not of the left ventricle Recent studies of cyanotic patients have, however, clearly shown that the myocardium of the left ventricle is not seriously injured when the oxygen saturation of the arterial blood is far lower than that of normal venous blood Therefore it seems unlikely that the scarring of the myocardium is due to the low oxygen content of the venous blood Gasul and Loeffler[8] in 1949 suggested that the low pressure in the pulmonary artery after birth was the factor of prime importance in the failure of the left ventricle to receive adequate nutrition when the left coronary artery arose from the pulmonary artery This theory has been widely accepted, it offers a clear explanation of the striking difference in the symptoms produced by the anomalous origin of the right and left coronary arteries Recent evidence indicates that St. John Brooks postulation of the retrograde flow of blood through the anomalous coronary artery is a factor of great importance The thin walled right ventricle apparently can receive adequate circulation when supplied with blood under pulmonary pressure hence there is no stimulus for the development of collateral circulation The thick walled left ventricle, however, requires a high pressure in the coronary artery in order to receive adequate nutrition Unfortunately, as the nutrition becomes inadequate, the development of and the increase in the anastomoses between the two coronary arteries cause a retrograde flow of blood through the left coronary artery which arises from the pulmonary artery, consequently the development of collateral circulation further impairs the nutrition of the myocardium of the left ventricle Certainly it is the anomalous origin of the left descending coronary artery from the pulmonary artery which is of clinical importance Furthermore, this anomaly produces a distinctive clinical syndrome

NATURE OF THE MALFORMATION

This malformation affects the blood supply to the myocardium All four chambers of the heart are normally formed but the left coronary artery is mis-

placed Instead of arising in the normal fashion from the base of the aorta, it arises from the base of the pulmonary artery Consequently the left ventricle receives venous blood under low pressure Furthermore, if there are any anastomoses between the two coronaries, the blood flows from the right coronary artery to the left and a retrograde flow of blood is established through the left coronary artery, thus depriving the myocardium of nutrition (see below) Such anastomoses behave as arteriovenous aneurysms and tend to increase in size and number as the infant grows The nutrition of the myocardium of the left ventricle is seriously impaired The left ventricle becomes weakened and greatly dilated, owing to its poor nutrition, hypertrophy cannot occur It becomes a thin walled, greatly dilated chamber (see Figure XXIX-1) The myocardium is severely injured, and extremely scarred, calcification may occur in the large areas of infarction (see Figure XXIX-2 A)

As in all cases of failure of one side of the heart, there is back pressure on the other side, hence the healthy myocardium of the right ventricle hypertrophies By the time of death the left ventricle is a thin walled, markedly dilated chamber, its myocardium is greatly scarred and its endocardium is thickened, whereas the right ventricle is slightly hypertrophied and the myocardium is normal (see Figure XXIX-2 B)

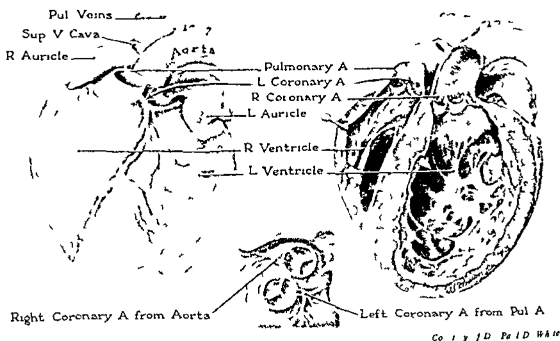


FIGURE XXIX-1 Anomalous origin of the left coronary artery from the pulmonary artery Infant

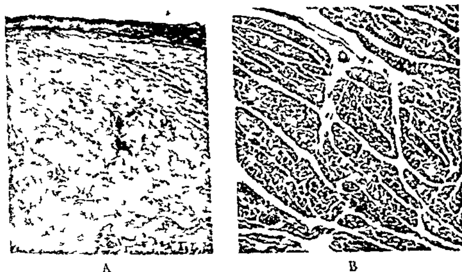


FIGURE XXIX–2 Anomalous origin of the left coronary artery from the pulmonary artery (A) Anterior wall of the left ventricle (x 35) showing infarct and calcification and (B) right ventricle (x 55) showing normal myocardium

COURSE OF THE CIRCULATION

During fetal life the lungs do not function and most of the blood in the pulmonary artery is pumped through the ductus arteriosus to the descending aorta Consequently the right ventricle pumps against systemic pressure and during fetal life the pressures in the aorta and the pulmonary artery are equal Furthermore, there is little or no difference in the oxygen content of the blood in the pulmonary artery and the aorta Therefore the anomalous origin of the coronary artery from the pulmonary artery has no effect upon the nutrition of the fetal myocardium. At birth the heart is normal in size and shape

After birth the only effect upon the circulation is that the left ventricle is nourished by venous blood under low pressure This alters the nutrition of the myocardium but does not affect the course of the circulation The course of the circulation is normal

PHYSIOLOGY OF THE MALFORMATION

The origin of the left coronary artery from the pulmonary artery has a twofold effect upon the circulation of the blood to the myocardium One is that the left ventricle is supplied with venous blood under low pressure The other results from the anastomoses which develop between the two coronary arteries The right coronary artery, which arises normally, receives blood under systemic

pressure, whereas the left coronary artery, which arises from the pulmonary artery, receives blood under pulmonary pressure Consequently, as Brooks[1] and more recently Edwards[9] have pointed out, as anastomoses develop between the left and right coronary arteries, the difference in pressure between the two arteries causes blood to flow from the right coronary artery into the left coronary and, furthermore, blood continues to flow in a retrograde direction through the left coronary to the pulmonary artery Thus blood is actually syphoned away from the left ventricular myocardium Both factors seriously impair the nutrition of the left ventricle It becomes greatly dilated and its wall abnormally thin Consequently the left ventricle is unable to maintain the systemic circulation

The hemodynamics of the circulation are altered only by the progressive weakening of the myocardium, which leads to progressive cardiac failure and death at an early age

CLINICAL FINDINGS

The baby appears to be perfectly normal at birth and usually remains so the first four to six weeks of life

Polypnea is often the outstanding complaint

Difficulty in feeding is frequently the complaint with which the infant is brought to the doctor This difficulty may be due to the general debility which results from the failing circulation or it may be due to the fact that the digestion of food precipitates pain

Pain and *symptoms of shock* which occur after feeding may be outstanding clinical symptoms The infant may draw up his legs and cry out with pain or he may suffer from episodes of circulatory collapse and shock During such an attack, the infant becomes limp and ashen in color, he may sweat profusely The pulse is barely palpable The attacks are at first infrequent and of brief duration but become progressively more severe and more frequent The picture is one of shock and is similar to that seen in an adult with an acute myocardial infarction

Episodes of collapse are occasionally the first manifestation

CARDIAC FINDINGS

The heart is usually enlarged by the time the infant is brought to the doctor The *heart sounds* are of *poor quality* and there may be a well marked *gallop rhythm* Nevertheless, since there is no gross structural abnormality of the heart, murmurs and thrills are strikingly absent

Cardiac failure inevitably follows The infant develops rapid respirations,

rales in the lungs, engorgement of the liver, and edema of the extremities Digitalis may be of temporary benefit but death from cardiac failure is inevitable unless an operation is developed which will enable the myocardium of the left ventricle to receive adequate nutrition

The clinical picture resembles that of cardiac failure due to an acute myocarditis, to an endocardial fibro-elastosis, or to some unknown cause, far more closely than it does the usual picture of a congenital malformation of the heart Not infrequently the doctor is at a loss to know whether or not there is any cardiac abnormality This is not surprising when it is recalled that the underlying pathology is not caused by any structural abnormality but by the faulty nutrition of the myocardium of the left ventricle

X RAY AND FLUOROSCOPIC FINDINGS

The heart is greatly enlarged, the enlargement is primarily caused by the great dilatation of the left ventricle In the anterior posterior position the heart is seen to be proportionally more enlarged to the left than to the right (see Figure XXIX-3) In the left anterior-oblique position, the right ventricle appears slightly enlarged and the huge left ventricle extends far posteriorly (see Figures XXIX-4 and 5) Great rotation of the infant is required for the left ventricle to clear the spinal column



FIGURE XXIX-3 Anomalous origin of the left coronary artery from the pulmonary artery (same patient as in Figure XXIX-4) Infant

pressure, whereas the left coronary artery, which arises from the pulmonary artery, receives blood under pulmonary pressure Consequently, as Brooks[1] and more recently Edwards[9] have pointed out, as anastomoses develop between the left and right coronary arteries, the difference in pressure between the two arteries causes blood to flow from the right coronary artery into the left coronary and, furthermore, blood continues to flow in a retrograde direction through the left coronary to the pulmonary artery Thus blood is actually syphoned away from the left ventricular myocardium Both factors seriously impair the nutrition of the left ventricle It becomes greatly dilated and its wall abnormally thin Consequently the left ventricle is unable to maintain the systemic circulation

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Episodes of collapse are occasionally the first manifestation

CARDIAC FINDINGS

The heart is usually enlarged by the time the infant is brought to the doctor The *heart sounds* are of *poor quality* and there may be a well marked *gallop rhythm* Nevertheless, since there is no gross structural abnormality of the heart, murmurs and thrills are strikingly absent

Cardiac failure inevitably follows The infant develops rapid respirations,

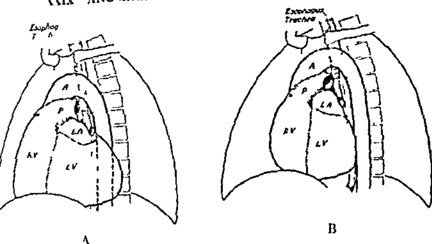


FIGURE XXIX-5 (A) Anomalous origin of the left coronary artery from the pulmonary artery and a large left ventricle and (B) normal heart Left anterior-oblique position Infant

The left ventricle is so greatly dilated that the right auricle and the right ventricle are displaced to the right and the left auricle is displaced upward and to the left. Consequently, upon delineation of the esophagus with a radio-opaque mixture, there is lateral and backward displacement of the esophagus by the left auricle (see Figure XXIX-4) The extent to which these chambers may be displaced is clearly demonstrated by angiocardiography, as shown in Figure XXIX-7

Upon fluoroscopy, a difference in the force of the contraction of the two ventricles should be sought When the infant is viewed in the left anterior oblique position, it may be possible to see normal pulsations along the anterior margin of the cardiac silhouette, but those along the posterior margin are weak or absent *

ELECTROCARDIOGRAPHIC FINDINGS

. . . · Bland

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drome and the electrocardiographic findings to those of acute myocardial infarction These authors reported a case in which the electrocardiogram showed inversion of T waves in all three standard leads and low voltage curves According to Keith et al,[10] the occurrence of a deep Q wave combined with inversion

* The author has also been able to distinguish the reverse phenomenon namely, vigorous pulsations over the left ventricle but no pulsations over the right ventricle in an infant with an abscess of the anterior mediastinum



Left anterior-oblique position

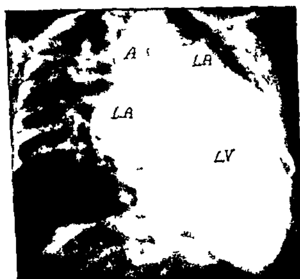


Right anterior-oblique position

FIGURE XXIX-4 Anomalous origin of the left coronary artery from the *pulmonary artery (same patient as in Figure XXIX-3) Infant*



Dextrogram



Levogram

FIGURE XXIX-7 Anomalous origin of the left coronary artery from the pulmonary artery Infant

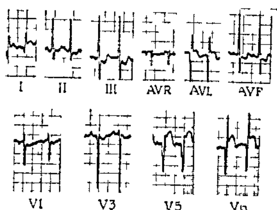


Figure xxix-6 Anomalous origin of the left coronary artery from the pulmonary artery Infant

of the T wave in Lead I and in AVL is the rule In addition, V_5 and V_6 usually show a deep Q wave followed by elevation of the S–T segment the T waves in these leads are frequently biphasic or inverted The T waves in Leads II and III may be upright and the voltage is usually normal

Inasmuch as the enlargement of the left ventricle is due to dilatation, not hypertrophy, in spite of the huge size of the left ventricle the electrocardiogram shows no evidence of left ventricular hypertrophy

The classic finding is that of a conspicuous Q wave and an inversion of the T wave in Lead I and in AVL combined with a deep Q wave and a marked elevation of the S–T segment in V_5 and V_6, as shown in Figure xxix–6 Such changes, when occurring in infancy, are nearly pathognomonic of the anomalous origin of the left coronary artery from the pulmonary artery The electrocardiogram is always abnormal but the above mentioned specific changes do not always occur

Digitalis may cause temporary improvement in the electrocardiogram, but the changes soon recur and thereafter persist There is no recovery from an anomalous coronary

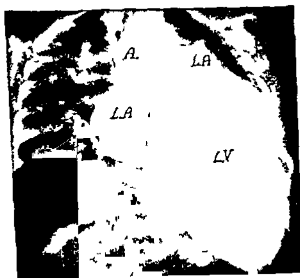
SPECIAL TESTS

Cardiac catheterization is of no diagnostic aid

Angiocardiography offers strong confirmatory evidence, as the left ventricle is an enormously dilated chamber with an extremely thin wall Since the contractions are weak, the dye lingers for a long time in this greatly dilated, thin walled left ventricle and there is no change in the size of the ventricle between systole and diastole The left ventricle is so huge that the right auricle and the right ventricle are displaced to the right and even the left auricle is pushed backward and upward (see Figure xxix–7) Its enormous size and thin wall are in



Dextrogram



Levogram

FIGURE XXIX-7 Anomalous origin of the left coronary artery from the pulmonary artery Infant

striking contrast to the small size and thick wall of the left ventricle seen in aortic stenosis or glycogen storage disease or even to the moderately thick walled left ventricle which occurs in endocardial fibro-elastosis

Retrograde aortography has been suggested[11] as of possible diagnostic value as only a single coronary artery (the right) would be seen to arise from the base of the aorta Negative evidence is never as satisfactory as positive evidence Indeed, even when both coronary arteries arise normally, it is not always possible to demonstrate them by aortography Therefore, failure to do so does not prove that the origin of the left coronary artery is abnormal

DIAGNOSIS

The outstanding findings in this malformation are great cardiac enlargement and cardiac failure with no murmurs and no cyanosis The failure is primarily left sided cardiac failure with râles in the lungs, usually there is also engorgement of the liver and there may be edema of the extremities Upon fluoroscopy the enlargement of the heart is seen to be due mainly to the increased size of the left ventricle The electrocardiogram is almost pathognomonic In Lead 1 it shows a deep Q wave and inversion of the T wave and in the unipolar precordial leads it shows changes characteristic of an acute anterior myocardial infarction The electrocardiogram is frequently said to look like that of an old man Moreover, day after day, week after week, the electrocardiographic changes characteristic of a recent acute infarction persist There is no recovery, hence there is no evidence of a healing infarction

DIFFERENTIAL DIAGNOSIS

The anomalous origin of the left coronary artery from the pulmonary artery must be differentiated from other conditions which cause severe injury to the left ventricular myocardium, cardiac enlargement, and cardiac failure in the absence of cyanosis or cardiac murmurs Therefore, the condition must be differentiated from severe anemia, hypertension, endocardial fibro-elastosis, Fiedler's myocarditis, vitamin deficiencies, glycogen storage disease of the myocardium, calcification of the coronary arteries, and occasionally from aortic stenosis and from a large ventricular septal defect, and in rare instances from acute rheumatic fever

Anemia when severe and of long duration, may cause enormous cardiac enlargement and cardiac failure The determination of the height of the hemoglobin indicates to what extent this factor may be operative If the anemia is

severe, an immediate effort should be made to raise the hemoglobin Repeated small blood transfusions are of great benefit

Essential hypertension[1] has been shown to be a cause of cardiac enlargement and cardiac failure in young children In the experience of the author, these children usually live longer than infants with an anomalous origin of the left coronary artery Infants with essential hypertension may suffer from severe cardiac failure but not from episodes of shock and collapse of the systemic circulation Determination of the blood pressure establishes the existence of hypertension. Infants with an anomalous origin of the left coronary artery from the pulmonary artery do not suffer from hypertension The electrocardiographic changes in the unipolar precordial leads are diagnostic.

Endocardial fibro-elastosis also causes difficulty in infancy and leads to cardiac enlargement and cardiac failure with a gallop rhythm but without endocardial murmurs The electrocardiogram shows evidence of left ventricular hypertrophy and inversion of the T waves in V_5 and V_6 Angiocardiography shows a rounded ventricular cavity which is not greatly dilated, with little change in its size between systole and diastole (see Section c)

Myocarditis of unknown etiology is relatively common in infants between seven and fifteen months of age The condition is manifested by the relatively sudden onset of cardiac failure in a previously healthy infant Respirations are rapid and the liver is engorged The heart is enlarged, the sounds are of poor quality A soft systolic murmur may occasionally be present or there may be no murmur, a well marked gallop rhythm is audible The heart is dilated A variety of electrocardiographic changes have been reported but the electrocardiogram does not show evidence of hypertrophy of either ventricle Only when the inflammation is extremely extensive are the curves of low voltage The T waves may be of low amplitude or inverted As the infant improves the electrocardiogram returns to normal

The response to digitalis is often gratifying The infant regains compensation, his appetite improves, and he appears normal Generally it requires a period of six to twelve months for the heart sounds to return to normal Nevertheless, many infants eventually make an excellent recovery The condition may be the same as *Fiedler s myocarditis* in which there is round cell infiltration of the myocardium In a number of instances, however, the infant makes an apparently complete recovery, therefore it is impossible to determine the pathology

Glycogen storage disease of the cardiomuscular type causes great cardiac enlargement in early infancy The heart sounds are of poor quality and a gallop

rhythm is common but endocardial murmurs are absent The infant is markedly hypotonic and severely retarded By five months of age the electrocardiogram shows a short P–R interval and a slight prolongation of the QRS interval (see Section D)

Vitamin deficiencies, especially *deficiency of vitamin B,* have been shown by Waring[13] to be a possible cause of cardiac failure in infancy Vitamin B deficiency is associated with right sided cardiac enlargement, whereas an anomalous origin of the left coronary artery causes left sided enlargement Studies concerning the vitamin B excretion and the vitamin B stores of the body should establish or exclude this factor

Calcification of the coronary arteries is an extremely rare condition, which leads to cardiac failure and death in early infancy Evidence of abnormal calcification elsewhere or evidence of abnormal calcium metabolism may suggest the diagnosis *In the only case which the author has ever seen, the heart was not* greatly enlarged nor was the wall of the left ventricle abnormally thin

Aortic stenosis, when severe in early infancy, may show electrocardiographic changes similar to those produced by an anomalous coronary artery In case of doubt, angiocardiography differentiates the two conditions

The Eisenmenger complex may occasionally be confused with an anomalous coronary artery The author has seen one infant who had an inconstant systolic murmur and a gallop rhythm combined with electrocardiographic changes suggestive of an anomalous left coronary artery, the baby also suffered from attacks of pallor and exhaustion, during which time she drew up her legs as if in pain *Eventually this infant, with the aid of digitalis, regained compensation* and presented the clinical picture of a patient with a large ventricular septal defect and increased pulmonary blood flow

Acute rheumatic fever without endocardial involvement may occur in early infancy but such an infection is extremely rare When it does occur, the infant is obviously acutely ill with high fever, which is in contrast to the findings in an infant with an anomalous coronary artery arising from the pulmonary artery In rheumatic fever the electrocardiogram frequently shows a *prolongation of* the P–R interval and only rarely is there inversion of the T waves in Lead I The author has never seen changes in the unipolar precordial leads which resembled those of acute anterior myocardial infarction in infants with acute rheumatic fever

Cardiac enlargement of unknown etiology, or so-called "idiopathic hypertrophy," frequently leads to cardiac failure It is from this group that all the

above mentioned conditions, including the anomalous origin of the left coronary artery from the pulmonary artery, have been differentiated Clinical diagnosis of the anomalous origin of the left coronary artery from the pulmonary artery is made by the characteristic electrocardiographic finding of a deep Q wave and inversion of the T waves in Lead I combined with evidence in the unipolar pre cordial leads of an acute anterior myocardial infarction which never heals

TREATMENT

Digitalis may be of temporary benefit Digitalis, however, does not increase the pressure in the pulmonary artery and hence cannot greatly improve the blood supply to the left ventricle The improvement is but temporary

Recently many ideas have been advanced for the surgical treatment of this condition. Beck and Brofman[14] have suggested that the coronary artery be anastomosed to the carotid artery so that it can receive arterial blood under systemic pressure. Edwards[9] has advocated ligation of the anomalous left coronary artery

cardium of the left ventricle.

Paul and Robbins[15] were the first to report the injection of talc into the peri cardium in order to stimulate the development of collateral circulation, in a manner similar to that advocated for coronary thrombosis For an infant who is severely ill, this procedure has the advantage that it is simple and can be per formed rapidly

Sabiston[16] has operated on a number of patients in whom the left coronary artery arose anomalously from the pulmonary artery He has found a striking difference in color between the two ventricles The right ventricle has a normal reddish color, whereas the left ventricle is yellowish The first baby died as the chest was opened and the diagnosis was confirmed at autopsy Although the second baby was in severe cardiac failure at the time of operation Sabiston was able to paint the left ventricle with phenol and then suture the left lung over the left ventricle in an effort to establish collateral circulation to the myocardium This baby survived the procedure and slowly regained compensation At fifteen months of age although the heart was greatly enlarged, the infant had learned to walk and the mother could see no difference between her baby and any other infant. At five and a half years of age this child was still well

In 1939 Sabiston[17] operated on an infant in whom the diagnosis of the anom alous origin of the left coronary artery was made at two and a half months In

this infant Sabiston found a localized aneurysm in the myocardium and conclusively demonstrated the retrograde circulation of blood through the left coronary artery by obtaining fully oxygenated bright red blood from the left coronary artery at its base Therefore, in addition to painting the myocardium with phenol, he ligated the left coronary artery

Poudrage of the myocardium with or without ligation of the anomalous coronary artery offers real hope Ligation of the anomalous coronary artery is indicated if retrograde flow through the coronary artery can be demonstrated If, however, anastomoses between the two coronary arteries have not been established, ligation of the left descending coronary artery cuts off what little blood supply is available to the left ventricle and may be immediately fatal Therefore the oxygen content of the blood in the anomalous coronary should be determined, or its color compared with that of the blood in the pulmonary artery, prior to ligation of this vessel

For surgical treatment to be really effective, the diagnosis must be made early and operation performed before the myocardium has been severely damaged Prompt operation is imperative because most babies die within four months after the onset of symptoms

PROGNOSIS

The prognosis is poor Although in rare instances adequate collateral circulation may develop early and thus give the individual a normal life span, the vast majority of infants suffer from progressive weakening of the myocardium of the left ventricle and death occurs between three and five months of age Occasionally an infant may live for ten or twelve months Early operation offers real hope for the prevention of injury to the left ventricular myocardium If such proves to be possible, the prognosis will be changed from hopeless to good

SUMMARY

The anomalous origin of the left coronary artery from the pulmonary artery causes difficulty because the thick walled left ventricle is supplied with venous blood under low pressure and, furthermore, as anastomoses develop between the right and the left coronary artery, blood is syphoned away from the left ventricle The myocardium is weakened and destroyed There is great dilatation of the left ventricle, which becomes extremely thin walled The condition causes progressive cardiac enlargement and cardiac failure with rales in the lungs and engorgement of the liver Inasmuch as murmurs and thrills are absent, the condition more closely resembles a myocarditis than a malformation of the heart

The electrocardiogram is of great diagnostic aid, the unipolar precordial leads show the pattern of acute myocardial infarction The electrocardiogram looks like that of an old man

Angiocardiography shows an enormous left ventricle with an extremely thin wall, in which the dye lingers for a long time

The condition requires differentiation from other causes of cardiac failure in which murmurs are absent namely, severe anemia, hypertension, endocardial fibro-elastosis, Fiedler's myocarditis, glycogen storage disease, and occasionally from aortic stenosis, a large ventricular septal defect, and acute rheumatic fever The electrocardiographic changes differentiate the anomalous origin of the coronary artery from these conditions

Numerous operations have been suggested to improve the circulation to the myocardium The most effective treatment so far devised is poudrage of the myocardium combined with ligation of the anomalous coronary artery at its base, provided retrograde flow of blood in that coronary artery can be demonstrated. In order to gain maximum benefit from treatment, operation should be performed at the earliest possible moment so as to prevent irreparable injury to the myocardium

The prognosis is poor Most infants die between three and five months of age. Operation may yet completely alter the prognosis

B *Coronary Arteriovenous Fistulae*

Coronary arteriovenous fistulae are comparatively rare Steinberg, Baldwin, and Dotter,[18] in their extensive review of the subject, expressed the opinion that such fistulae were rarer than tumors of the heart As in most malformations, the first such cases were found at autopsy As our diagnostic acumen increases, a steadily increasing number of these aneurysms are being diagnosed during life

Coronary aneurysms may be mycotic in origin, that is, secondary to infection and emboli, or due to rupture of a vessel secondary to arteriosclerosis, or they may be congenital in origin It is the last mentioned group with which the following discussion is concerned A review of the embryology of the coronary vessels will help to clarify the nature of the anomaly

EMBRYOLOGY

Grant[19] was the first to observe an aneurysm in the myocardium of an infant with a malformed heart This led him to study the embryology of the coronary vessels In his beautiful study of the development of coronary vessels in the rabbit heart,[illegible] he clearly shows the nature of this malformation His work may be

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EMBRYOLOGY

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briefly summarized as follows In the tiny embryo the coronary veins appear before the coronary arteries as outpouchings from the left horn of the sinus venosus and spread around the right and left atrioventricular grooves and also through the dorsal interventricular furrow to the apex of the heart From these regions the veins grow out over the surface of the ventricles, pierce the myocardium, and connect with the interventricular spaces At a later date the coronary arteries appear as endothelial sprouts from the base of the aorta and unite with the capillary network and with the intertrabecular spaces Thereafter, as the coronary arteries and veins grow and the myocardium condenses, the intertrabecular spaces are reduced to capillaries and are incorporated into the arteries and veins to form an integral part of the adult coronary circulation Thus the capillary bed of the myocardium is composed of elements from the coronary vessels and from the intertrabecular spaces Normally these spaces retain their connection with the interior of the ventricle as the so-called thebesian vessels

From these studies Grant concluded that the curious aneurysm in the myocardium of the infant's heart represented a persistence of the intertrabecular sinuses which normally exist between the muscle columns of the embryonic heart He offered this theory as an explanation of the blood filled sinusoidal network through which the coronary vessels communicated with the cavity of the ventricle

These studies clearly show the origin and the fundamental nature of coronary arteriovenous fistulae and in the author's opinion they also indicate how it is possible that occasionally one of the coronary arteries, although normally formed, fails to connect with its coronary vein and instead opens directly into the auricle or the ventricle Therefore it seems to the author that these two anomalies of the coronary vessels are closely related

In the former some of the intertrabecular spaces fail to become incorporated into the capillary bed and a coronary arteriovenous fistula results, in the latter the coronary artery fails to connect with the intertrabecular spaces and opens directly into one of the chambers of the heart

NATURE OF THE MALFORMATION

A coronary arteriovenous fistula is formed when one or more branches of the coronary vessels connect with the intertrabecular spaces but these spaces fail to become incorporated into the capillary bed When this occurs, the coronary vessels and the intertrabecular spaces form dilated, tortuous loops which become adherent to one another Thus the fistula is composed of fusiform vessels which

become elongated and the dilated, tortuous loops become coiled in a serpentine fashion and are firmly adherent to one another The location, number, and size of these fistulae vary from case to case

In most instances the tortuous vessels are relatively large but the opening into the auricle or the ventricle is small Occasionally these channels open into the coronary sinus The small size of the opening into the ventricle undoubtedly raises the pressure in the aneurysmal loops and may cause them to increase in number and size, in rare instances rupture of the aneurysm occurs

In the other type of coronary abnormality, the coronary artery appears to develop normally but fails to connect with the capillaries in the usual manner, instead it opens directly into the right auricle or the right ventricle Under such circumstances the wall of the artery is normal and, although there is an arteriovenous connection, it is small Indeed, it is smaller than a normal ductus arteriosus The condition places a constant load on the heart, it is, however usually so slight that it causes no symptoms

COURSE OF THE CIRCULATION

The course of the circulation is altered only by the volume of blood which flows through the coronary circulation Nevertheless since the shunt is from the aorta to an area of low pressure, it is usually of considerable magnitude Furthermore, arterial blood is shunted into the right auricle or the right ventricle and therefore the oxygen content of the blood directed to the lungs may be abnormally high

PHYSIOLOGY OF THE MALFORMATION

The physiology of the malformation is essentially that of an arteriovenous aneurysm Some of the blood from the coronary arteries flows into the dilated sinusoidal spaces, which in turn open into the heart As previously mentioned, the openings into the capillaries and into the thebesian vessels are small, hence the sinusoidal spaces tend to become progressively enlarged

The arteriovenous fistulae increase the work of the left ventricle and also decrease its nutrition Hence the condition may lead to cardiac failure

When the coronary artery is normally formed but opens directly into the right auricle or right ventricle instead of connecting with the coronary veins, it forms an arterial pathway from the aorta to the right side of the heart The physiology is similar to that of other arteriovenous aneurysms but, inasmuch as the vessel is relatively small (far smaller than a normal ductus) and the wall of

the vessel is normal, the fistula does not tend to increase in size as the patient grows, for this reason it is usually a benign lesion

CLINICAL FINDINGS

The clinical symptoms are remarkably few in both types of anomalies The majority of these children are asymptomatic and the condition is discovered on a routine physical examination

Dyspnea on exertion may occur in older patients, as the heart begins to fail *Signs of congestive cardiac failure* may eventually develop

CARDIAC FINDINGS

The heart may or may not be enlarged

A continuous murmur and a thrill are characteristic of this anomaly Although the location of the murmur and the thrill varies with the location of the fistulae, they are usually maximal over the lower portion of the precordium Both the murmur and the thrill appear to be superficial in origin Neill and Mounsey[21] have emphasized that the murmur is loud in mid systole and then wanes toward the second heart sound, and again increases in intensity in mid diastole and again wanes

X RAY AND FLUOROSCOPIC FINDINGS

There may be no abnormality visible in the x ray or there may be evidence of cardiac enlargement (see Figure XXIX-8) In rare instances an aneurysmal dilatation may be demonstrated Scott[22] reported a case in which the aneurysmal mass was so large that it caused backward displacement of the esophagus Intracardiac calcification was demonstrated in a case by Colbeck and Shaw[23] Such findings are, however, the exception, not the rule

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram is usually not remarkable unless the condition progresses to left ventricular hypertrophy and cardiac failure Knoblich and Rawson[24] reported a case in which the electrocardiogram suggested the occurrence of an acute posterior myocardial infarction with extension into the septum These authors suggested that, when such an electrocardiographic pattern persisted over a period of weeks without evidence of healing, the rare possibility of a coronary arteriovenous fistula should be considered

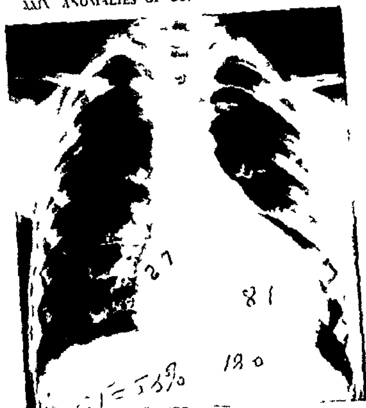


FIGURE XXIX–8 Coronary arteriovenous fistula (same patient as in Figures XXIX–9 12) Child

SPECIAL TESTS

Cardiac catheterization usually shows an increase in the oxygen content of the blood in the right auricle or in the right ventricle and thereby indicates the chamber into which the fistula or coronary artery opens

Angiocardiography usually demonstrates the tortuous vessels in the myocar [illegible]

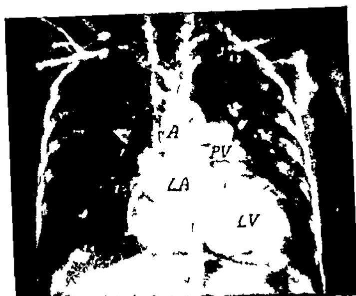
[illegible] tricle, dye will be seen to enter the right side of the heart immediately after the aorta has filled Frequently a pathway can be traced from the aorta to the right auricle or the right ventricle, as shown in Figures XXIX–10 and 11

Aortography also readily demonstrates both conditions by the opacification of the anomalous pathway which arises from the aorta Figure XXIX–12 shows a coronary arteriovenous fistula and Figure XXIX–13 shows a coronary artery opening into the right auricle





FIGURE XXIX–9 Coronary arteriovenous fistula (same patient as in Figure XXIX–8) Child



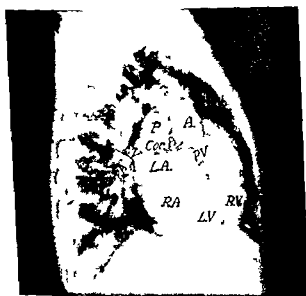


FIGURE XXIX–10 Coronary arteriovenous fistula (in diastole) Child





FIGURE XXIX-9 Coronary arteriovenous fistula (same patient as in Figure XXIX-8) Child



FIGURE XXIX-12 Coronary arteriovenous fistula (same patient as in Figure XXIX-8) Child

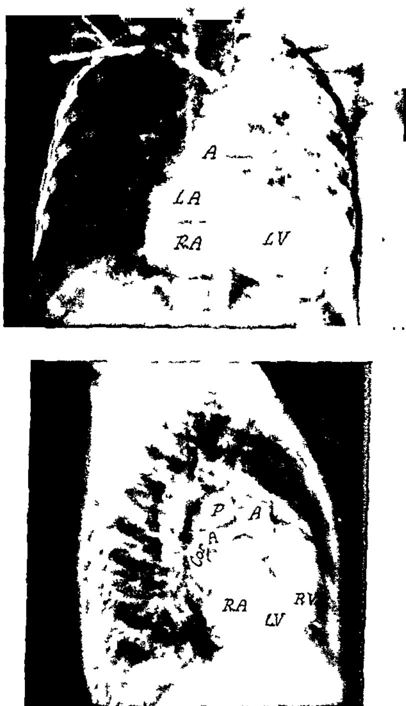


FIGURE XXIX-11 Coronary arteriovenous fistula (in systole) Child

The coronary arteriovenous fistula and the coronary artery which opens directly into the right side of the heart can be differentiated from each other by angiocardiography or aortography

TREATMENT

Many patients are asymptomatic and require no treatment Successful surgical ligation of the anomalous vessel has now been repeatedly performed Gasul et al.[29] have emphasized that corrective surgery is relatively easy and does not require hypothermia or the use of a cardiopulmonary by pass Hence, if there is evidence of cardiac strain or progression of symptoms, operation is indicated

PROGNOSIS

The prognosis is better than one would expect Steinberg, Baldwin, and Dotter[5] emphasized the fact that nine out of thirteen autopsy reports were of patients over fifty three years of age, and that two of the nine were over seventy and two others were eighty years of age With the recent interest in diagnosis and surgery, a number of patients have been operated upon in childhood or early adolescence

SUMMARY

Coronary arteriovenous fistulae and an anomalous branch of a coronary artery which opens directly into one of the chambers of the heart are being reported with increasing frequency

Both conditions represent failures of the coronary artery to connect with the coronary veins in the normal manner An arteriovenous fistula or aneurysm is formed when the coronary artery fails to connect with the coronary vein and the intertrabecular sinusoidal spaces persist When a branch of the coronary artery opens directly into the right side of the heart, no aneurysm forms, but the basic physiology and the clinical manifestations are similar

The course of the circulation is altered only by the volume of blood shunted from the aorta to the lesser circulation

The physiology affects the nutrition of the myocardium

The clinical symptoms are remarkably few in childhood The condition, if severe may lead to congestive cardiac failure later in life

The outstanding cardiac finding is a superficial continuous murmur and thrill over the lower part of the precordium

The electrocardiogram is usually not of diagnostic significance Nevertheless,

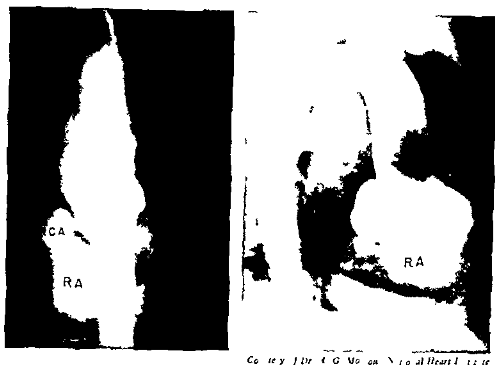


FIGURE XXIX-13 Coronary artery opening into the right auricle. Child

Coronary arteriography, as developed by Dotter and Frische,[5] probably offers the best means for a precise localization of the abnormality

DIAGNOSIS

The diagnosis is based upon the occurrence of a continuous murmur and a thrill over the precordium, both are superficial and are usually maximal over the lower precordium The diagnosis is confirmed by angiocardiography or aortography

DIFFERENTIAL DIAGNOSIS

These two conditions require differentiation from a patent ductus arteriosus and other conditions which cause a continuous murmur

Patency of the ductus arteriosus differs from the abnormalities under discussion in that in a patent ductus the thrill, although readily palpable, is not superficial and both the murmur and the thrill are maximal over the base of the heart

The differentiation of other conditions which cause a continuous murmur, such as a rupture of an aneurysm of the sinus of Valsalva into the lesser circulation, an arteriovenous aneurysm in the thoracic wall, and occasionally a pulmonary arteriovenous aneurysm and a hemi truncus arteriosus, are discussed in detail in Chapter XXV

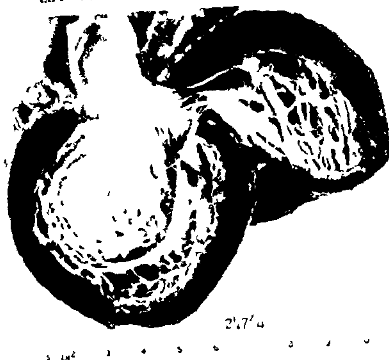


FIGURE XXIX-14 Endocardial fibro-elastosis Infant

there is an excess of elastic tissue and of collagenous material [9] through which nothing seems to penetrate The layer of material varies from one to several millimeters in thickness The author remembers one case in which the inner half of the left ventricular wall was replaced by such abnormal tissue

The abnormality may involve any or all the chambers of the heart It occurs frequently in combination with valvular abnormalities, especially in combination with congenital anomalies of the mitral and aortic valves—notably mitral insufficiency of the type in which the chordae tendineae are abnormally short and the valve is grossly deformed It is, however a striking fact that basically the heart is normally formed and there are no abnormal communications between the two sides of the heart Although Thomas et al [9] have reported a series of cases in children and young adults, the condition occurs primarily in young infants Furthermore, when the left ventricle is the chamber which is primarily involved the condition produces a distinctive clinical syndrome It is with this condition that the following discussion is concerned

COURSE OF THE CIRCULATION

Inasmuch as there are no abnormal communications between the two sides of the heart the course of the circulation is normal

the finding of persistence of the signs of an acute posterior myocardial *infarction* should suggest the possibility of this congenital abnormality

Angiocardiography or aortography clinches the diagnosis of a *coronary* arteriovenous fistula and aids in the diagnosis of communication between a branch of the coronary artery and the right auricle or the right ventricle Coronary arteriography may show the exact anatomical location of the abnormality

The diagnosis is based on the occurrence of a continuous murmur and thrill over the lower portion of the precordium in an asymptomatic patient

The condition requires differentiation from a patency of the ductus arteriosus, from a rupture of an aneurysm of the sinus of Valsalva into the right side of the heart, and from the other conditions from which these malformations must be differentiated

Surgical correction is possible if the condition causes cardiac embarrassment

The prognosis in most instances is remarkably good and may be rendered excellent by surgery

C *Endocardial Fibro elastosis*

Endocardial fibro-elastosis is generally believed to be congenital in origin The condition is known by various names endocardial fibro-elastosis, fibro-elastosis, subendocardial fibro-elastosis, or subendocardial sclerosis The term endocardial dysplasia was suggested by Prior and Wyatt,[27] as they believe it is a developmental disorder of the mesenchymal tissue

ETIOLOGY

The etiology is unknown The condition occurs in association with valvular abnormalities and also as an isolated anomaly It was formerly regarded as secondary to myocarditis and to round cell infiltration of the myocardium In most instances, however, there is no evidence of inflammation Indeed, the absence of an inflammatory reaction has been one of the strong arguments in favor of the theory that the condition is congenital in origin Furthermore, it may occur in more than one offspring of the same parentage The incidence also varies in different parts of the country, it is remarkably high in Buffalo, New York

NATURE OF THE MALFORMATION

The abnormality concerns the endocardium The heart itself is normally formed The endocardium is white and appears to be thickened but it is smooth and glistening (see Figure XXIX-14) Immediately beneath the endocardium



FIGURE XXIX–15 Endocardial fibro-elastosis Infant

shadow show weak and wavy contractions or "snake like" movements rather than the normal strong, lateral contractions

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram usually shows a balanced axis and, as Lambert et al[20] have pointed out, the unipolar precordial leads show evidence of left ventricular hypertrophy In V_5 and V_6 the R wave is tall and the T wave is inverted (see Figure XXIX–16)

SPECIAL TESTS

Cardiac catheterization is of no aid in diagnosis

Angiocardiography shows a normal circulation and no evidence of a shunt but the dye lingers for a long time in the left ventricle, the cavity of the left ventricle appears rounded and there is little change between systole and diastole The wall of the left ventricle is slightly thicker than normal but it is not as greatly thickened as it is in glycogen storage disease of the myocardium (compare Figure XXIX–17 with Figure XXIX 21)

DIAGNOSIS

The diagnosis is based upon the finding of cardiac enlargement and cardiac failure, a gallop rhythm, and the absence of endocardial murmurs, combined with electrocardiographic evidence of left ventricular hypertrophy in V_5 and V_6 It is confirmed by angiocardiographic evidence of the absence of significant

PHYSIOLOGY OF THE MALFORMATION

The condition affects the contractility and the nutrition of the myocardium The myocardium is unable to contract normally The heart becomes dilated and the myocardium slightly hypertrophied and there is strikingly little change in the size or shape of the left ventricular chamber between systole and diastole In addition, it seems inevitable that the thebesian vessels are obliterated and the nutrition of the inner layer of the myocardium is thereby impaired

CLINICAL FINDINGS

The infant appears normal at birth Sometime between six weeks and six months of age he becomes irritable and fails to thrive

Cyanosis and clubbing are absent, as there is no intracardiac communication

Respirations become rapid *Rales* in the lungs may develop

The liver enlarges and frequently extends to the umbilicus

Vomiting may occur with decompensation

Edema of the extremities is a late manifestation of cardiac failure

CARDIAC FINDINGS

The heart is markedly enlarged The apex beat is palpable in the anterior axillary line The heart sounds are of *poor quality,* there is usually a *gallop rhythm* but endocardial murmurs are strikingly absent

Cardiac failure usually occurs between seven and ten months of age Indeed, it is the development of cardiac failure with rapid respirations, engorgement of the liver, and edema of the extremities which most frequently brings the infant to the doctor

X RAY AND FLUOROSCOPIC FINDINGS

The heart is enlarged to the right and the left There may be fullness of the pulmonary conus The vascularity of the lungs appears normal (see Figure XXIX-15)

Examination in the left anterior oblique position shows that the left ventricle is greatly enlarged It requires considerable rotation of the infant for the left ventricle to clear the spinal column In the right anterior-oblique position a barium swallow reveals backward displacement of the left auricle This is usually associated with lateral displacement of the esophagus in the anterior posterior view

Fluoroscopy reveals that the pulsations along the margin of the cardiac



FIGURE XXIX–15 Endocardial fibro-elastosis Infant

shadow show weak and wavy contractions or snake like movements rather than the normal strong, lateral contractions

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram usually shows a balanced axis and, as Lambert et al[30] have pointed out, the unipolar precordial leads show evidence of left ventricular hypertrophy In V_5 and V_6 the R wave is tall and the T wave is inverted (see Figure xxix–16)

SPECIAL TESTS

Cardiac catheterization is of no aid in diagnosis

Angiocardiography shows a normal circulation and no evidence of a shunt it the dye lingers for a long time in the left ventricle, the cavity of the left ven icle appears rounded and there is little change between systole and diastole he wall of the left ventricle is slightly thicker than normal but it is not as reatly thickened as it is in glycogen storage disease of the myocardium (com are Figure xxix–17 with Figure xxix–21)

DIAGNOSIS

The diagnosis is based upon the finding of cardiac enlargement and cardiac failure a gallop rhythm, and the absence of endocardial murmurs, combined with electrocardiographic evidence of left ventricular hypertrophy in V_5 and V_6 It is confirmed by angiocardiographic evidence of the absence of significant

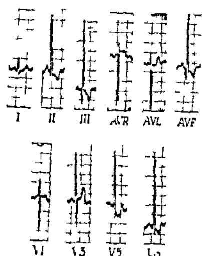


FIGURE XXIX-16 Endocardial fibro-elastosis Infant

changes in the size and shape of the cavity of the left ventricle in systole and diastole

DIFFERENTIAL DIAGNOSIS

The condition requires differentiation from myocarditis, anomalous origin of the left coronary artery from the pulmonary artery, and glycogen storage disease

Myocarditis is by far the most difficult condition to diagnose with certainty Although a myocarditis may progress to intractable cardiac failure and death, usually the infant responds to digitalis and over the months he gradually improves The electrocardiographic findings are variable but changing, they improve as the infant improves A myocarditis is a diffuse process and there is usually no evidence of either right or left ventricular hypertrophy When the myocarditis is extremely severe, the electrocardiographic curves may be of low voltage

Anomalous origin of the left coronary artery from the pulmonary artery shows similar evidence of cardiac enlargement and absence of endocardial murmurs The electrocardiogram, however, shows the pattern of an acute anterior myocardial infarction but no evidence of left ventricular hypertrophy

Glycogen storage disease which affects the heart also causes great cardiac enlargement and little change in the size of the ventricle between systole and diastole The hypotonia of the infant, combined with the electrocardiographic finding of a P–R interval of approximately the same length as the QRS interval, sharply differentiates it from an endocardial fibro-elastosis



Dextrogram



Levogram

FIGURE XXIX–17 Endocardial fibro-elastosis Infant

ATMENT

Digitalis should be given in full doses Diuretics are indicated if there is ema.

Any operation to increase the circulation to the myocardium is strongly indi ted Paul and Robbins[15] were among the first to advocate the application of cle to the myocardium

Sabiston[16] has tried the removal of the anterior portion of the pericardium and the application of phenol to the myocardium In at least two instances such treatment has resulted in marked improvement

Without operation the condition is usually fatal

PROGNOSIS

The prognosis is poor Most infants die between seven and twelve months of age A few die at even younger ages The fact that the condition occasionally occurs in children and young adults indicates that there may be variation in the severity of the endocardial fibro-elastosis and that at times it may be relatively mild or increase slowly It is, of course, possible that in some instances it is the end result of an acquired condition Operation may greatly improve the prognosis

SUMMARY

Endocardial fibro-elastosis is a condition in which there is thickening of the subendocardial layer of the heart, especially of the left ventricle, which is believed to be congenital in origin and may be inheritable

The condition leads to weakening of the inner wall of the myocardium with progressive cardiac enlargement A gallop rhythm is common but murmurs and thrills are absent Cardiac failure usually occurs between the seventh and tenth months of life

Fluoroscopy shows diffuse cardiac enlargement, the left auricle may be slightly enlarged, the vascularity of the lungs is normal and contractions are poor

The electrocardiogram shows evidence of left ventricular hypertrophy and inversion of the T waves in V_5 and V_6

Angiocardiography shows little change in the size of the left ventricular cavity between systole and diastole

Treatment is directed toward the improvement of the circulation and the nutrition of the myocardium

Digitalis is indicated if the heart is enlarged and diuretics should be given if there is evidence of failure

Surgical measures to increase the vascularization of the myocardium may be of great benefit

Without operation the prognosis is poor Most infants die between seven months and one year of age

D *Glycogen Storage Disease*

Glycogen storage disease of the heart and striated muscles is a widespread metabolic disease which also affects the central nervous system [31]

The etiology is unknown It appears to be a familial disease Nadas[37] has reported three siblings to be affected in one family and we have cared for one family with two infants affected with the disease A history of consanguinity is relatively frequently obtainable.

NATURE OF THE MALFORMATION

The heart is normally formed The defect in the glycogen metabolism is such that glycogen in large quantities is deposited in the muscles, especially in the cardiac muscle The heart muscle becomes swollen with glycogen and the walls of the ventricles are greatly thickened The individual fibers are greatly enlarged and filled with glycogen but their contractile power is weakened[33] The tremendous size of the heart and the thickness of the ventricular wall are contrasted with the heart of an infant of the same age in Figure XXIX–18

The abnormal deposition of glycogen is not limited to the cardiac muscle It is found in all the striated muscles and may even be present in the smooth muscles. Oppenheimer[31] has shown that the central nervous system is also affected



A B

FIGURE XXIX–18 (A) Glycogen storage disease and (B) normal heart. Infants of same age

COURSE OF THE CIRCULATION

The course of the circulation is normal

PHYSIOLOGY OF THE MALFORMATION

The physiology of the disease concerns the metabolism of glycogen For a discussion of this aspect of the disease the reader is referred to the authorities in this field such as Cori [34] The hemodynamics of the circulation are normal

CLINICAL FINDINGS

The baby appears normal at birth but *difficulty in nursing* soon becomes apparent and steadily increases The infant may have a *feeble cry* even at birth

Cyanosis is absent, as there is no shunt Circumoral cyanosis is, however, frequently noted, it results from the sluggish circulation

Growth and development are severely retarded During the first month growth is slow and thereafter it virtually ceases The infant fails to hold up his head at the normal time and even fails to follow a light with his eyes

The tongue is also loaded with glycogen Consequently it becomes enlarged The *macroglossia* and the *mental retardation* may suggest that the infant is a cretin

Hypotonia is marked The muscles feel reasonably firm but they have no strength

Respirations become rapid *Respiratory infections* and pneumonia are common, as the infant's resistance to disease is poor *Râles* in the lungs are of frequent occurrence

The liver may or may not be enlarged

The stools may have an odor of a "bakery," that is, a strong odor of yeast

CARDIAC FINDINGS

The heart is greatly enlarged It may even be enlarged at birth The heart action is so feeble that the apex thrust may not be palpable The heart sounds are of *poor quality* A *gallop rhythm* is the rule but endocardial murmurs are absent

X RAY AND FLUOROSCOPIC FINDINGS

X ray and fluoroscopy reveal diffuse cardiac enlargement, especially of the ventricles, and the pulsations are strikingly weak The vascularity is normal (see Figure XXIX–19) The esophagram usually reveals left auricular enlargement



FIGURE XXIX–19 Glycogen storage disease Infant

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram shows a remarkably short P–R interval and a QRS which is slightly prolonged, so that the P–R interval equals the QRS interval[10] (see Figure XXIX–20) The deflections are usually high and may be extraordinarily high In addition the T waves in the standard leads may be inverted and there may be depression of the S–T segments The changes in the P–R interval which are so characteristic of this condition become more pronounced as the infant becomes older They were not present in an infant who had marked cardiomegaly when he was seen at one month of age and who died at two months of age and at autopsy proved to have the disease The changes in the electrocardiogram are however, usually present by five months of age

SPECIAL TESTS

The red blood cell count the *hemoglobin* and the *hematocrit* may show evidence of anemia

Analysis of the *glycogen content* of the *red blood cells* shows it to be abnormally high

Muscle biopsy and analysis of the *glycogen content* of the muscle establishes the diagnosis

Angiocardiography is seldom necessary to differentiate the condition from other causes of cardiac enlargement It does, however, offer confirmatory evi

COURSE OF THE CIRCULATION

The course of the circulation is normal

PHYSIOLOGY OF THE MALFORMATION

The physiology of the disease concerns the metabolism of glycogen For a discussion of this aspect of the disease the reader is referred to the authorities in this field such as Cori [34] The hemodynamics of the circulation are normal

CLINICAL FINDINGS

The baby appears normal at birth but *difficulty in nursing* soon becomes apparent and steadily increases The infant may have a *feeble cry* even at birth

Cyanosis is absent, as there is no shunt Circumoral cyanosis is, however, frequently noted, it results from the sluggish circulation

Growth and development are severely retarded During the first month growth is slow and thereafter it virtually ceases The infant fails to hold up his head at the normal time and even fails to follow a light with his eyes

The tongue is also loaded with glycogen Consequently it becomes enlarged The *macroglossia* and the *mental retardation* may suggest that the infant is a cretin

Hypotonia is marked The muscles feel reasonably firm but they have no strength

Respirations become rapid *Respiratory infections* and pneumonia are common, as the infant s resistance to disease is poor *Rales* in the lungs are of frequent occurrence

The liver may or may not be enlarged

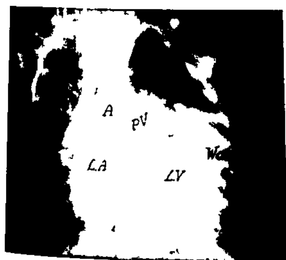
The stools may have an odor of a 'bakery,' that is, a strong odor of yeast

CARDIAC FINDINGS

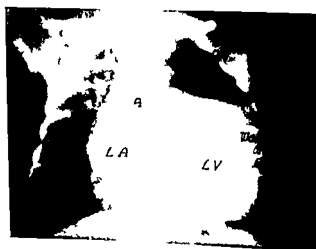
The heart is greatly enlarged It may even be enlarged at birth The heart action is so feeble that the apex thrust may not be palpable The heart sounds are of *poor quality* A *gallop rhythm* is the rule but endocardial murmurs are absent

X RAY AND FLUOROSCOPIC FINDINGS

X ray and fluoroscopy reveal diffuse cardiac enlargement, especially of the ventricles, and the pulsations are strikingly weak The vascularity is normal (see Figure XXIX-19) The esophagram usually reveals left auricular enlargement



Ventricular systole



Ventricular diastole

FIGURE XXIX–21 Glycogen storage disease. Infant

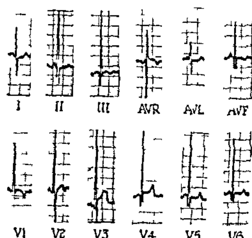


FIGURE XXIX-20 Glycogen storage disease. Infant

dence in that it shows that the left ventricle is abnormally thick and the cardiac contractility poor and that there is little change in the size of the ventricle between systole and diastole (see Figure XXIX-21)

DIAGNOSIS

The diagnosis is to be suspected in a markedly hypotonic infant in whom the heart is enormously enlarged, the heart sounds are of poor quality, and endocardial murmurs are absent The electrocardiogram shows wide deflection and an extremely short P–R interval which approximately equals the duration of the QRS complex

The diagnosis is confirmed by muscle biopsy and the demonstration of an abnormally high glycogen content in the muscle

DIFFERENTIAL DIAGNOSIS

The condition may be confused with congenital amyotonia or even with cretinism The great cardiac enlargement combined with the absence of endocardial murmurs calls for differentiation from the anomalous origin of the left coronary artery from the pulmonary artery, from endocardial fibro-elastosis, and from acute myocarditis

An anomalous origin of the left coronary artery from the pulmonary artery causes totally different changes in the electrocardiogram Lead I usually shows a deep Q and elevation of the S–T segment and the unipolar precordial leads show the pattern of an acute anterior myocardial infarction

Fluoroscopy shows poor contraction of the ventricles

The electrocardiogram shows a short P-R interval and a very slight prolongation of the QRS interval and may show tall deflections and inversion of the T waves

Angiocardiography shows a thick walled left ventricle and little change in the size or shape of the heart during systole and diastole

The diagnosis is to be suspected when a markedly hypotonic infant suffers from great cardiac enlargement, a gallop rhythm, and no endocardial murmurs The diagnosis is confirmed by muscle biopsy

Treatment is directed toward correction of the metabolic disturbance

Digitalis is indicated to improve heart action

The prognosis is poor Most infants die from bulbar paralysis or from cardiac failure between five and seven months of age

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Endocardial fibro elastosis probably more closely resembles glycogen storage disease than does any of the other conditions This is especially true in young infants before the characteristic electrocardiogram has developed and when the difference in the thickness of the ventricular wall in the two conditions is not great Indeed, it is the hypotonia of the infant which gives the clue to the diagnosis the two conditions are seldom confused as the disease progresses A baby with endocardial fibro elastosis does not show the extreme hypotonia which occurs in glycogen storage disease Furthermore, the electrocardiogram becomes markedly different

Myocarditis usually occurs abruptly in a previously healthy child There is no history of poor weight gain, of slow development, or of marked hypotonia The electrocardiogram is subject to variation but does not show the short P–R interval characteristic of glycogen storage disease

TREATMENT

Treatment must be directed to the correction of the metabolic disease Therein lies the real hope for cure

Digitalis may help to tide the infant over until his metabolism can be corrected

Antibiotics may protect him from respiratory and pulmonary infections

PROGNOSIS

The prognosis is poor Until the metabolic disease can be corrected, the condition progresses Death quite as often results from bulbar paralysis[31] as from cardiac failure Most of these infants die between five and seven months of age A few infants die at two months and a few children have lived to four years

SUMMARY

Glycogen storage disease, which involves the heart and muscles of the body, is a metabolic disturbance which is congenital in origin, appears to be familial, and may be associated with consanguinity in the parents

The disease involves all the muscles of the body and also the central nervous system The infant is markedly hypotonic and severely retarded Growth is extremely slow, great difficulty is encountered in feeding, as there is real difficulty in swallowing and difficulty in metabolizing the food which is ingested

The heart is greatly enlarged A gallop rhythm is common Endocardial murmurs and thrills are absent The liver may be enlarged

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Such an anomaly frequently occurs when the abnormality involves the left ventricle as well as the mitral valve

Congenital mitral stenosis most commonly occurs in association with malformations of the left side of the heart, notably defective development of the left ventricle, endocardial fibro-elastosis, valvular and subvalvular aortic stenosis, and coarctation of the aorta When the left ventricle is defective, the mitral valve may be atretic but more commonly the mitral orifice and the mitral valve are abnormally small The chordae tendineae are short Occasionally the mitral ring is normal in size and the valve itself is stenotic A mitral ring of normal size occurs more frequently when the left ventricle is normal than when the left ventricle is defective Consequently pure mitral stenosis may occur as an isolated malformation and be the cause of severe obstruction to the pulmonary circulation The mitral leaflets are thickened and fused together and the chordae tendineae may be abnormally short Occasionally, however, the mitral orifice may be abnormally small and the mitral valve may be thickened, yet the chordae tendineae may be relatively long, as in the case reported by Ferencz et al [1]

Mitral stenosis and insufficiency frequently occur together The author has twice seen this abnormality of the mitral valve in combination with coarctation of the aorta In some instances the mitral ring is normal in size but the leaflets are fused together and hang down into the left ventricle like a bag with an opening in its bottom The margin of the orifice is rolled and hence the valve leaflets are unable to open, so that there is mitral stenosis as well as mitral insufficiency (see Figure XXX–1)

This condition is to be distinguished from Ebstein s anomaly on the left side of the heart which occasionally occurs in combination with a corrected transposition In Ebstein s anomaly the septal leaflet of the tricuspid valve is plastered against the wall of the septum and a new orifice is formed deep within the ventricle In the anomaly under discussion the valve arises normally from the atrioventricular ring but the outer margins of the leaflets are fused together and therefore unable to open normally Thus the valve is both stenotic and insufficient

COURSE OF THE CIRCULATION

During fetal life the occurrence of either mitral insufficiency or mitral [illegible] no-

[illegible] cle

[illegible] the pressure in the left auricle and thereby decreases the flow of blood from the right auricle to the left auricle consequently during fetal life the left

CHAPTER XXX

MALFORMATIONS OF THE LEFT AURICLE

MALFORMATIONS of the left auricle are comparatively rare, there are, however, a variety of such abnormalities The *mitral valve* may be either insufficient or stenosed or both Occasionally, a membrane is formed across the left auricle, dividing it into a large upper chamber and a small lower chamber, this condition is known as the *triauricular* heart In rare instances the musculature of the left auricle is so defective that the wall balloons out, forming an *aneurysmal dilatation* of the left chamber

Valvular abnormalities are discussed in Section A Section B concerns the triauricular heart, and Section C illustrates the findings in aneurysmal dilatation of the left auricle

A *Anomalies of the Mitral Valve*

Congenital mitral insufficiency and congenital mitral stenosis both occur as isolated malformations or in association with other malformations Furthermore, either abnormality may occur separately or they may occur together The physiology and the clinical manifestations are closely similar to those of acquired valvular disease There is, however, no reason to believe that either is rheumatic or even infectious in origin

NATURE OF THE MALFORMATION

Congenital mitral insufficiency occurs when the endocardial cushions, from which the mitral valve develops, fail to grow and unite in the normal manner, a cleft remains between two of the leaflets Such is the nature of the mitral insufficiency which occurs in association with a defect of the ostium primum type A cleft mitral valve is also almost an integral part of a persistent ostium atrioventriculare commune Such a cleft may occasionally occur with a ventricular septal defect and also with a defect of the ostium secundum type Thus, by and large, a cleft in the mitral valve occurs in association with various types of septal defects

Unfortunately, in many instances, the nature of the insufficiency is far more serious The mitral orifice is distorted, the chordae tendineae are abnormally short, and the valve leaflets are plastered against the wall of the left ventricle

from the right auricle through the foramen ovale, hence there may be premature closure of the foramen ovale The course of the fetal circulation in mitral stenosis is shown in Figure XXX–2

At birth with the expansion of the lungs and the establishment of the pulmonary circulation, the blood is returned to the left auricle in the normal manner The difficulty in the expulsion of blood through the mitral orifice raises the pressure in the left auricle and the foramen ovale promptly closes Thereafter the course of the circulation is basically normal The course of the circulation in mitral stenosis is illustrated in Diagram XXX–1

PHYSIOLOGY OF THE MALFORMATION

The physiological changes caused by mitral insufficiency and mitral stenosis are due to the difficulty in the expulsion of blood from the left auricle They are

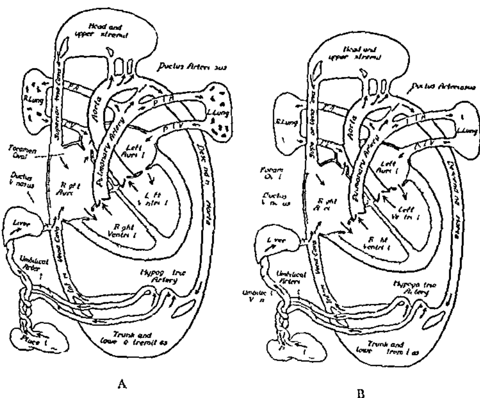


FIGURE XXX–2 Fetal circulation (A) Congenital mitral stenosis and (B) normal heart

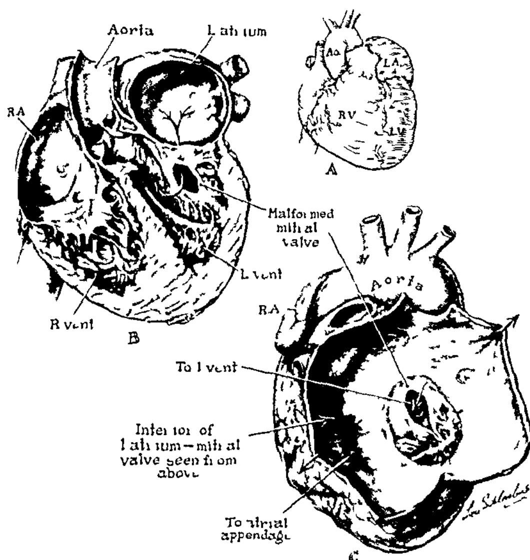


Figure XXX-1 Congenital mitral stenosis and insufficiency and enormous dilatation of the left auricle A coarctation of the aorta had been surgically corrected

side of the heart receives less than its normal quota of blood and the right ventricle pumps proportionally more The second effect of the high pressure in the left auricle is the back pressure it creates on the embryonic pulmonary vascular bed Ferencz and Dammann[2] have shown that the changes in the pulmonary vascular bed appear at an extremely early age in infants in whom the back pressure is effective during fetal life, that is, in congenital mitral stenosis and in a triauricular heart The high pressure in the left auricle lessens the flow of blood

from the right auricle through the foramen ovale, hence there may be prema ture closure of the foramen ovale The course of the fetal circulation in mitral stenosis is shown in Figure xxx-2

At birth with the expansion of the lungs and the establishment of the pul monary circulation, the blood is returned to the left auricle in the normal man ner The difficulty in the expulsion of blood through the mitral orifice raises the pressure in the left auricle and the foramen ovale promptly closes There after the course of the circulation is basically normal The course of the circula tion in mitral stenosis is illustrated in Diagram xxx-1

PHYSIOLOGY OF THE MALFORMATION

The physiological changes caused by mitral insufficiency and mitral stenosis are due to the difficulty in the expulsion of blood from the left auricle They are

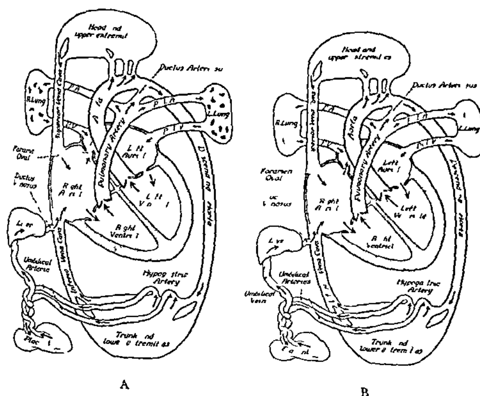
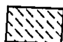


FIGURE XXX-2 Fetal circulation (A) Congenital mitral stenosis and (B) normal heart

Diagram XXX–1



Arterial blood (fully saturated)



Small admixture of venous blood
No visible cyanosis



Venous and arterial blood
Cyanosis visible

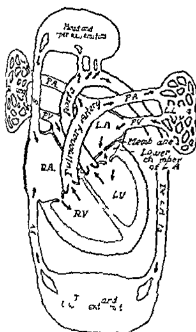


Venous blood

Diagram XXX–1

Congenital mitral stenosis

The essential feature in this malformation is the obstruction to the flow of blood from the left auricle to the left ventricle It matters little whether there is stenosis of the mitral valve or a diaphragm above the mitral valve with only a small perforation In either case there is difficulty in the expulsion of blood from the left auricle and consequently the left auricle becomes dilated and its wall hypertrophied The increased pressure in the left auricle is transmitted back into the pulmonary veins and thence through the capillaries to the arterial side consequently there is increased resistance in the pulmonary vascular bed and increased pressure in the pulmonary artery and in the right ventricle hence there is right ventricular hypertrophy



Triauricular heart

The circulation is fundamentally normal The blood from the right auricle flows into the right ventricle and is pumped out through the pulmonary artery to the lungs where it is oxygenated The oxygenated blood is returned to the left auricle thence it flows to the left ventricle and is pumped out through the aorta to the systemic circulation and returned by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again

Clinical diagnosis The early onset of dyspnea and pulmonary congestion especially when associated with brief attacks of syncope followed by lethargy are characteristic of obstruction to the flow of blood within the left auricle. The heart is enlarged there may or may not be a presystolic murmur a the apex Inasmuch as there is no shunt there is no cyanosis and no clubbing X ray and fluoroscopy reveal right sided cardiac enlargement fullness of the pulmonary conus increased vascular marking and also great enlargement of the left auricle. The electrocardiogram shows a right axis deviation and evidence of right ventricular hypertrophy

basically the same as those which occur in acquired mitral disease but they occur at a far earlier age. Indeed, they may be initiated during fetal life. The left auricle becomes greatly enlarged and the pressure in the left auricle rises.

Mitral insufficiency causes greater left auricular enlargement than does mitral stenosis because of the regurgitation of blood into the left auricle. Thus, with each auricular contraction, the left auricle is required to pump not only all the blood which it receives from the pulmonary veins but also the blood which is regurgitated from the left ventricle; consequently the left auricle undergoes both dilatation and hypertrophy. In contrast to this, mitral stenosis causes difficulty only in the ejection of blood from the left auricle. Although the work of the left auricle may be greatly increased, the volume of blood which it must pump is not increased; hypertrophy is greater than dilatation. Dilatation results only when the left auricle fails to empty itself.

In both conditions the pressure in the left auricle is increased. As the pressure in the left auricle rises, the pressure in the pulmonary veins also rises. This in turn obstructs the flow of blood through the capillaries and thereby the pulmonary vascular resistance is increased; hence there is pulmonary hypertension. Consequently the pressure in the pulmonary artery becomes elevated and the work of the right ventricle is increased. Thus there is not only left auricular enlargement but also pulmonary hypertension and, secondary to this, the work of the right side of the heart is increased. The right ventricle and the right auricle are dilated and hypertrophied.

CLINICAL FINDINGS

The clinical manifestations are similar to those of acquired mitral stenosis or insufficiency.

Cyanosis and *clubbing* are absent. There is no abnormal opening between the two sides of the heart; hence there is no possibility of a shunt. The oxygen saturation of the arterial blood is normal. Cyanosis results only from peripheral stasis and increased deoxygenation in the capillaries.

Left sided chest deformity develops early because of the enlargement of the right ventricle.

Dyspnea and *polypnea* develop early. Dyspnea, on exertion, becomes marked.

Cough is a common complaint. It may be caused by pulmonary congestion or may be secondary to pressure on the recurrent laryngeal nerve due to the great enlargement of the left auricle. The latter condition frequently causes a brassy cough.

Pulmonary congestion is common The infant frequently suffers from con gestion in the lungs, asthmatic bronchitis, and pneumonia, when there is mitral stenosis, he may suffer from attacks of acute pulmonary edema

The liver becomes engorged and frequently extends to the umbilicus

Edema is a late manifestation of cardiac failure Frequently the puffiness of the eyelids is noticed before the swelling of the feet becomes apparent

When congenital mitral stenosis is severe the condition may cause grave difficulty within the first year of life Usually the infant does well for a month or longer, depending on the severity of the mitral stenosis As the systemic blood flow becomes inadequate, the infant fails to thrive He becomes irritable and ceases to gain weight and grow normally

Attacks of sweating pallor and *pain* followed by *limpness*, which may progress to *loss of consciousness* are late manifestations of severe mitral stenosis[3] During the spell the *pulse* becomes imperceptible and the *heart rate* slows There may be transitory cyanosis The *attacks of syncope* are usually brief but are followed by *lethargy* for several hours Such attacks are of ominous prognostic import, as they are indicative of collapse of the systemic circulation

CARDIAC FINDINGS

The cardiac findings are similar to those of acquired mitral disease The heart undergoes progressive enlargement The enlargement concerns mainly the left auricle and the right side of the heart The right ventricle is both dilated and hypertrophied and the pulmonary artery is dilated The murmurs and thrills also are similar to those of acquired mitral valvular disease

The second heart sound over the pulmonary area is usually markedly accentuated because of the pulmonary hypertension

A harsh systolic murmur which is of maximal intensity at the apex and well transmitted to the axilla and back, is usually readily heard The murmur has the blowing quality of a murmur of mitral insufficiency In addition a *systolic thrill* is palpable at the apex

With the advent of *cardiac failure* the heart sounds become of poor quality There are, however, both a *systolic* and a *mid diastolic murmur* audible at the apex and frequently there is a *gallop rhythm*

A presystolic murmur and a thrill at the apex which are accentuated by exercise, are the most characteristic of all findings in congenital mitral stenosis Although this murmur is not always obvious at first, it can usually be detected in infants upon careful examination after exercise in the left lateral position

X RAY AND FLUOROSCOPIC FINDINGS

The heart is diffusely enlarged both to the right and to the left (*see Figure* XXX–3) When mitral insufficiency is extreme, the left auricle may project to the right of the sternum beyond the margin of the right auricle Indeed, it may be so huge and the pulsations along the margin of the shadow so marked that it is difficult to believe that the entire shadow is caused solely by the left auricle Under such circumstances the condition may be clarified by angiocardiography The pulmonary conus is full and the vascular markings are increased There may or may not be pulsations in the hilar vessels

Upon delineation of the esophagus with barium in the right anterior-oblique *position, the left auricle is usually seen to be greatly enlarged Occasionally the* displacement of the esophagus is better seen in the anterior posterior view than in the oblique view (see Figures XXX–3, 4, and 5)

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram usually shows a right axis deviation in the standard leads and evidence of marked right ventricular hypertrophy in the unipolar pre cordial leads The P waves are notched and frequently of increased duration, the total P–R interval may be prolonged (see Figure XXX–6)

SPECIAL TESTS

The red blood cell count the amount of available hemoglobin and *the hem*

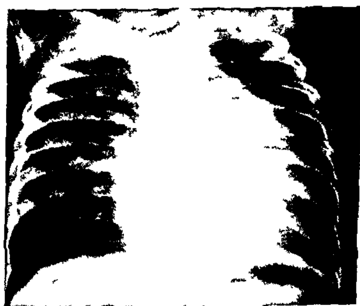


FIGURE XXX–3 Congenital mitral stenosis Infant



Left anterior-oblique position



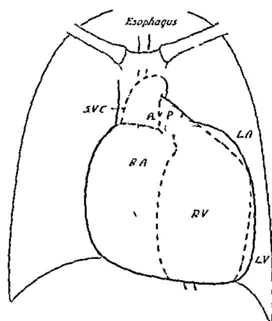
Right anterior-oblique position

FIGURE XXX-4 Congenital mitral stenosis Infant

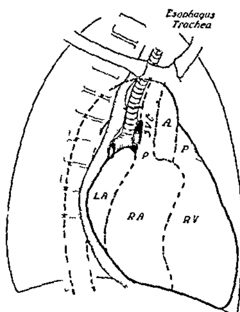
atocrit reading remain remarkably normal There is no tendency to develop polycythemia Nevertheless, in spite of the severe cardiac failure there is usually no anemia

Cardiac catheterization shows no evidence of a shunt but the pressure tracings reveal increased pressure in the right ventricle and in the pulmonary artery and an increased resistance in the pulmonary capillary bed The wedge pressure is elevated These findings are due to the increased pressure in the pulmonary veins, which results from the elevation of the pressure in the left auricle

Angiocardiography is of aid in the differentiation of mitral stenosis from mi

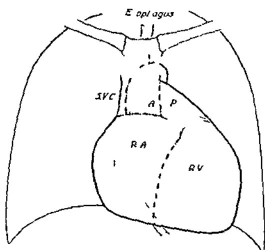


Anterior posterior position

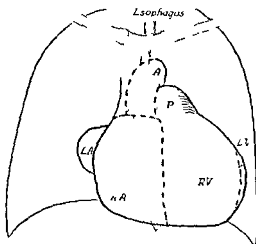


Right anterior-oblique position

ADULT



Anterior posterior position



Anterior posterior position

CHILD

FIGURE XXX-5 Large left auricle Adult and child

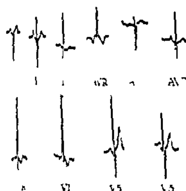


FIGURE XXX-6 Congenital mitral stenosis Infant

tral insufficiency In both abnormalities the right auricle and the right ventricle are enlarged and there is a huge left auricle in which the contrast substance lingers for a long time

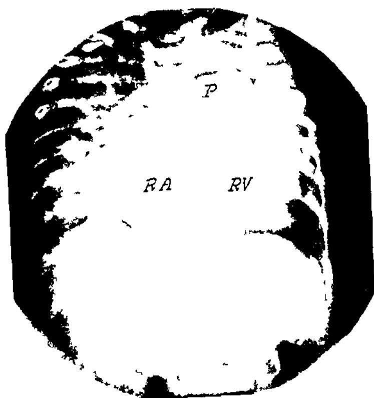
Mitral stenosis is differentiated from mitral insufficiency mainly by the ease with which the left ventricle fills When there is severe mitral stenosis, little dye passes into the left ventricle consequently the left ventricle is poorly visualized and the aorta may appear abnormally small, as shown in Figure xxx-7 Indeed, the left ventricle and the aorta may never be delineated In contrast to this, when there is mitral insufficiency the dye passes readily into the left ventricle Nevertheless, dye in relatively high concentration remains for a long time in the left auricle (see Figure xxx-8) the difference in the size of the left auricle in systole and diastole can usually be seen

DIAGNOSIS

The diagnosis is based upon the finding of mitral valvular disease in an infant or young child with marked cardiac enlargement and cardiac failure and no cyanosis or clubbing There may be a presystolic murmur at the apex or a systolic murmur which is well transmitted to the axilla The diagnosis is substantiated by the x ray evidence of enlargement of the left auricle and electrocardiographic evidence of right ventricular hypertrophy It is confirmed by the angiocardiographic demonstration of the large left auricle in which the dye lingers for a long period of time

DIFFERENTIAL DIAGNOSIS

Both conditions require differentiation from acquired mitral disease and occasionally from primary pulmonary hypertension and from a triauricular heart



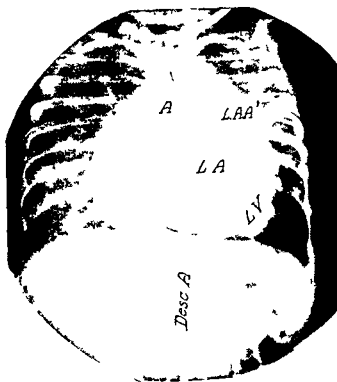


FIGURE XXX-7 Congenital mitral stenosis Infant

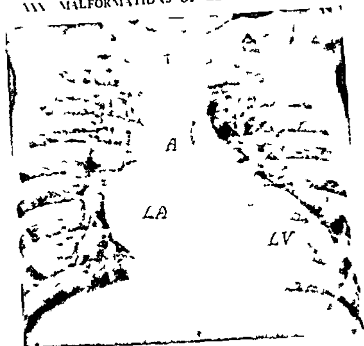


FIGURE XXX-8 Congenital mitral insufficiency with coarctation of the aorta

Rheumatic heart disease with mitral stenosis or insufficiency produces similar auscultatory findings The clue to the diagnosis of congenital mitral stenosis is given by the early age at which the mitral disease is manifested, combined with the absence of any history or evidence of a rheumatic infection or indeed of any other illness The sedimentation rate remains normal the red blood cell count also remains normal There is no anemia in spite of long standing chronic cardiac failure

Primary pulmonary hypertension causes accentuation of the pulmonary second sound and right ventricular hypertrophy but should not be confused with abnormalities of the mitral valve as there is no presystolic murmur and no great left auricular enlargement

A triauricular heart (see below) also causes difficulty in the expulsion of blood from the left auricle There is, however, no suggestion of a presystolic murmur or thrill Furthermore, the malformation usually causes difficulty at an even earlier age than does congenital mitral stenosis

COMMONLY ASSOCIATED MALFORMATIONS

The malformations which commonly occur in conjunction with congenital mitral stenosis are coarctation of the aorta endocardial fibro-elastosis, and a gross defect in the auricular septum of the ostium secundum type

Congenital mitral insufficiency is frequently associated with a defect of the ostium primum type, it is part and parcel of persistent ostium atrioventriculare commune and may occur with a ventricular septal defect

Coarctation of the aorta is frequently associated with other left sided cardiac lesions When it occurs with congenital mitral stenosis or insufficiency, each produces its own characteristic findings There is high pressure in the upper extremities and the femoral pulses are weak or absent The heart is enlarged and there is evidence of mitral valvular disease The absence of other evidence of rheumatic fever and the knowledge that the condition dates from an early age indicate that the valvular disease is congenital The coarctation of the aorta places an additional load on the heart This may be corrected by surgery but the correction of the valvular abnormality is far more difficult Unfortunately it is the valvular abnormality which usually causes the major difficulty

Endocardial fibro elastosis also occurs in combination with congenital mitral stenosis It, too, lessens the efficiency of the heart It, too, is difficult to alleviate by surgery

A gross defect in the auricular septum of the ostium secundum type combined with mitral stenosis constitutes Lutembacher s syndrome The defect in the auricular septum relieves the strain on the mitral valve and totally alters the clinical findings (see Chapter XXIII, Section B)

An ostium primum defect becomes a distinctive clinical entity when there is a cleft in the mitral valve Thus it is the occurrence of mitral insufficiency which completes the clinical picture (see Chapter XXIII, Section C)

A persistent ostium atrioventriculare commune is usually associated with an abnormality of the mitral and tricuspid valves These may fuse together and both may be competent but frequently one or both of the valves have a cleft in the septal leaflet (see Chapter XXIII, Section D)

Ventricular septal defects are occasionally complicated by mitral insufficiency Under such circumstances, in addition to the harsh systolic murmur caused by the ventricular defect, there is a harsh systolic murmur at the apex which is well transmitted to the axilla and there is also evidence of enormous enlargement of the left auricle

TREATMENT

The surgical correction either of congenital mitral stenosis or of congenital mitral insufficiency is quite different from that of acquired mitral stenosis or insufficiency

Although mitral commissurotomy is easily performed and of great benefit to a patient with acquired mitral stenosis, a congenitally malformed valve is frequently so severely deformed that commissurotomy is not possible Furthermore, the condition may be associated with endocardial fibro-elastosis of the left ventricle Nevertheless, occasionally the structure of the valve is such that commissurotomy is possible, as in the case reported by Braudo et al[4] Dr Henry Bahnson has operated on two infants with severe mitral stenosis in whom he was able to increase the size of the mitral orifice Therefore, inasmuch as the prognosis is hopeless without operation, surgical intervention is indicated if the infant is in difficulty

The ease with which a mitral insufficiency can be corrected also depends upon the structure of the mitral valve It is virtually impossible to reconstruct a small valve which is plastered against the wall of the left ventricle If, however, the mitral insufficiency is caused by a cleft in the mitral valve, it is relatively easy to correct. Kirklin and others have found that it is usually possible to suture the clefts of the leaflets together and thereby to overcome the insufficiency

PROGNOSIS

Congenital mitral insufficiency and stenosis both carry a serious prognosis, as both lead to progressive cardiac enlargement and pulmonary hypertension Congenital mitral insufficiency is usually not as serious as is congenital mitral stenosis furthermore, if the condition is caused by a cleft in the mitral valve, it can be corrected by surgery When there is congenital mitral stenosis, unless the mitral orifice can be increased in size the condition leads to progressive cardiac enlargement and to ever increasing changes in the pulmonary vascular bed Cardiac failure occurs early The child rarely survives beyond three years of age

There are, however, all grades of severity If the mitral stenosis is relatively mild, the condition may be compatible with life for a number of years The author has seen congenital mitral stenosis more frequently in association with coarctation of the aorta than as an isolated abnormality Under such circumstances it is the severity of the mitral stenosis, not the coarctation of the aorta, which limits the patient s life

SUMMARY

Congenital mitral stenosis and insufficiency are rare anomalies Either may occur as an isolated malformation or in combination with other malformations Both malformations cause difficulty in the emptying of the left auricle, and

consequently cause increased pressure in the lungs and on the right side of the heart in a manner similar to that of acquired mitral valvular disease There is no abnormal communication between the two sides of the heart, hence there is no shunt When congenital mitral stenosis is severe, the infant usually does well for the first few weeks, then he becomes irritable and fails to gain and grow normally He develops a cough and dyspnea and may suffer from attacks of pain associated with pallor and a feeble pulse The attacks may progress to syncope followed by several hours of lethargy

The heart becomes enlarged, there are usually both a systolic and a mid diastolic murmur over the precordium and in cases of mitral stenosis there is a true crescendo presystolic murmur and thrill at the apex

The x ray shows great enlargement of the left auricle, enlargement of the right ventricle, and dilatation of the pulmonary artery

The electrocardiogram shows notching of the P waves, a right axis deviation, and evidence of right ventricular hypertrophy

Cardiac catheterization shows no shunt but reveals increased pressure in the right ventricle and in the pulmonary artery

Angiocardiography reveals a greatly enlarged left auricle in which the dye lingers for a long time If there is severe mitral stenosis, little dye passes into the left ventricle and the left ventricle and aorta are poorly visualized, whereas if there is mitral insufficiency, the left ventricle fills promptly and dye remains in both the left auricle and the left ventricle through several cardiac cycles

Both conditions require differentiation from acquired valvular disease and occasionally from pulmonary hypertension and a triauricular heart

Congenital mitral stenosis is frequently complicated by other left sided cardiac lesions When it occurs with a gross defect in the auricular septum of the ostium secundum type, it constitutes Lutembacher s syndrome

Congenital mitral insufficiency completes the syndrome produced by an ostium primum defect, also occurs with a persistent ostium atrioventriculare commune, and may also occur with a ventricular septal defect

Surgical repair of a cleft mitral valve is relatively easy, whereas if the mitral valve is plastered against the wall of the left ventricle it may be impossible to correct the condition

Mitral stenosis is usually extremely difficult to relieve but occasionally the structure is such that mitral commissurotomy is possible

The prognosis is poor Congenital mitral stenosis usually leads to progressive cardiac enlargement and death before three years of age Congenital mitral in

sufficiency is more often amenable to treatment and hence the prognosis is far better for this malformation than for congenital mitral stenosis

B *Triauricular Heart*

A triauricular heart is a rare but well recognized pathological entity [6] In this malformation there are three auricles and two ventricles A brief review of the embryology will clarify the nature of the anomaly

EMBRYOLOGY

The lung buds form simultaneously with the heart and, as the pulmonary veins develop, their branches fuse together as they approach the heart Finally the main branches from the left and right lungs fuse together and a single pulmonary vein grows down to meet an outpouching from the left auricle [7] Normally as the left auricle expands, the first two branches, and later all four branches of the pulmonary vein become incorporated into the left auricle

In the malformation under discussion it appears that, although the outpouching from the left auricle expands normally to form a chamber into which the four pulmonary veins drain the orifice of the outpouching fails to expand and consequently a membrane persists which separates the upper portion of the left auricle from the lower portion of that chamber It is from the lower portion of the left auricle that the mitral valve opens into the left ventricle

Inasmuch as the pulmonary veins normally have been incorporated into the left auricle before the ostium secundum forms and furthermore, since in this anomaly the ostium secundum opens into the upper chamber, which is by far the larger of the two, it seems as if the anomaly must occur between Horizon XV when the pulmonary veins have developed and Horizon XVI when the ostium secundum forms that is, when the embryo is approximately one month of age

NATURE OF THE MALFORMATION

There are three auricles one right auricle and two left auricles The right auricle is normally formed The anomaly concerns only the double left auricle This auricle is divided into two chambers a large upper chamber into which all four pulmonary veins enter and a small lower chamber which leads to the left ventricle The upper chamber is separated from the right auricle by the interauricular septum Furthermore it is into this chamber that the ostium secundum opens The lower margin of the chamber is formed by a membrane which is pierced by a single small opening It is through this opening that blood can

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trickle into the tiny lower chamber from which the mitral valve opens into the left ventricle Figure xxx–9 is a drawing of such an abnormality

COURSE OF THE CIRCULATION

The course of the circulation is basically the same as that in congenital mitral stenosis (see Diagram xxx–1 and insert)

PHYSIOLOGY OF THE MALFORMATION

The physiological effect of this malformation is the same as that of congenital mitral stenosis, in that the tiny opening between the upper and the lower chambers *obstructs the flow of blood from the left auricle into the left ventricle* Consequently this anomaly elevates the pressure in the pulmonary veins and in the pulmonary capillary bed, this in turn increases the pressure in the pulmonary arterioles and the pulmonary arteries, and ultimately increases the work of the right ventricle

CLINICAL FINDINGS

The clinical findings are virtually identical with those of severe congenital mitral stenosis

The infant does well initially At some time between one and six months of age he ceases to thrive and fails to gain weight and to develop normally He becomes irritable

Attacks of sweating and *pallor* associated with a cry of *pain* are late manifestations of pulmonary venous obstruction The attacks may progress to *limpness* and *momentary loss of consciousness* During this time the *pulse* becomes imperceptible and the *heart rate* slows The *syncope* is usually brief but is often followed by a period of *lethargy* The attacks may be precipitated by feeding In one instance the mother was able to prevent spells by sitting the baby upright for his feeding

The attacks of syncope and *lethargy* are less readily recognized as cardiac in origin because murmurs are absent *Indeed, the attacks may be mistaken for petit mal*

CARDIAC FINDINGS

The heart is enlarged Its contour is similar to that found in congenital mitral stenosis The left auricle appears to be enlarged, as the upper chamber of the left

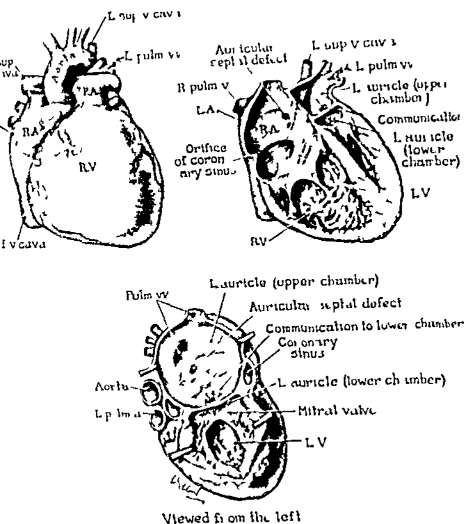


FIGURE XXX-9 Triauricular heart

auricle is greatly enlarged The right auricle, the right ventricle, and the pulmonary artery are also enlarged

There is, however, no presystolic murmur or thrill, as the blood trickles slowly and constantly from the upper chamber into the tiny lower chamber and thence through the mitral valve into the left ventricle Nevertheless, the absence of the presystolic murmur and thrill does not clinch the diagnosis, as a mitral stenosis can be so severe that no presystolic murmur can be elicited



Figure xxx-10 Triauricular heart Infant

X RAY AND FLUOROSCOPIC FINDINGS

The x ray and fluoroscopic findings may be identical with those of severe mitral stenosis, in some instances, however, the left auricular enlargement may not be visible in the anterior posterior view (see Figure xxx-10)

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram shows a right axis deviation in the standard leads and *evidence of right ventricular hypertrophy in the unipolar precordial leads*

DIAGNOSIS

The diagnosis may be difficult to make with certainty The finding of car diac enlargement, especially left auricular enlargement in the absence of mur murs, is characteristic of this condition Therefore, when an infant with evidence of great left auricular enlargement suffers from attacks of syncope followed by lethargy, such symptoms should suggest that the difficulty is caused by collapse of the systemic circulation, secondary to severe pulmonary venous obstruction

DIFFERENTIAL DIAGNOSIS

The condition requires differentiation from severe congenital mitral stenosis, from petit mal, and possibly from tumors of the left auricle

Mitral stenosis may be so severe that no murmur is audible Under such cir

cumstances it may be impossible to tell whether the obstruction lies at the mitral valve or at a slightly higher level Fortunately it makes no great difference, as the treatment is the same for both conditions

Petit mal may be considered because of the brief attacks of syncope followed by lethargy An electroencephalogram may aid in the diagnosis of epilepsy but the recording of a normal brain wave in an infant does not exclude the possibility of epilepsy The finding of cardiac enlargement, especially of the left auricle, suggests that the syncope is due to pulmonary venous obstruction If such is the case, angiocardiography will show that dye lingers for a long time in the left auricle

A tumor in the left auricle may give the same clinical findings as severe mitral stenosis, with the single exception that there is no evidence of valvular heart disease[8] The same is true of a patient with a triauricular heart The age of the patient should differentiate the two conditions, as tumors of the heart are rare in infancy and childhood and a patient with a triauricular heart seldom survives infancy

TREATMENT

Although to the author s knowledge operation has never been attempted, the structure of the left auricle is such as to indicate that under direct vision it might be possible to excise the entire membrane and thereby restore the heart to normal

PROGNOSIS

The prognosis is poor Most infants with a triauricular heart die at an early age The author, however, knows of two patients one of whom lived to nine years of age and the other to thirteen It is hoped that when the condition is diagnosed operation for the correction of the mitral obstruction will be attempted, as it appears possible that the anomaly might be corrected by surgery

SUMMARY

A triauricular heart is a rare anomaly in which the left auricle is divided into two chambers, a large upper chamber into which the pulmonary veins enter and a small lower chamber from which the mitral valve opens into the left ventricle The membrane between these two chambers is pierced by a tiny opening Consequently there is great difficulty in the expulsion of blood from the upper chamber into the left ventricle

The physiology is the same as that of congenital mitral stenosis The clinical and cardiac findings are essentially similar, except for the absence of a presystolic murmur and thrill

The malformation is of clinical importance because, if the condition were correctly diagnosed, it is quite possible that the membrane across the left auricle could be surgically removed and the circulation thereby restored to normal

C *Aneurysmal Dilatation of the Left Auricle*

Most cases of saccular dilatation of the left auricle are secondary to mitral insufficiency or mitral stenosis, they are usually the result of acquired disease The author has, however, studied one instance of congenital "aneurysmal" dilatation of the left auricle[9] This was in 1937

Illustrative Case

Case xxx–1 C C (Harriet Lane Home, No A–3887) Negro female First seen in 1937 at five years of age because of severe cardiac failure. The child preferred to lie on her right side and did so even when propped up in bed

The heart was markedly overactive and enormously enlarged There was extreme dextrorotation On percussion the cardiac dullness on the left extended to the mid clavicular line in the sixth left interspace and it extended equally far on the right The heart beat could be readily felt both to the right and to the left Indeed, it was so readily palpable on the left that some observers thought that the heart occupied its normal position

There was a harsh systolic murmur and thrill over the precordium in addition, both a systolic murmur and a low pitched mid diastolic murmur and a gallop rhythm were audible to the right of the sternum Fluoroscopic examination showed tremendous cardiac enlargement to the right and to the left, the cardiac pulsations were remarkably feeble There was no demonstrable displacement of the esophagus

The liver occupied its normal position and extended to the umbilicus There was no edema of the extremities

The electrocardiogram showed a rate of 245 per minute The P waves were of small amplitude and were inverted in Lead I and upright in Leads II and III At that time the use of precordial leads had not been developed

Autopsy No 15532 (performed by Dr James Semans) The heart was enormously enlarged The cardiac border extended 7 cm to the right and 10 cm to the left Upon opening the pericardium an incomplete dextrocardia was found The entire left side of the pericardial cavity was occupied by the left auricle The right ventricle lay to the extreme right, the left ventricle occupied a central position, and the apex pointed

downward and to the right (see Figure XXX-11) The great vessels were displaced to the right but lay in normal relation to each other

Further examination of the heart showed that the right auricle was slightly hypertrophied The foramen ovale was closed The right ventricle was dilated and hypertrophied The left auricle was huge On the posterior aspect of the left auricle there was a sharply demarcated saccular dilatation which measured 10 cm in its greatest diameter The wall of this sac was very thin and fibrous it contained only a few criss crossing muscle fibers

The left auricular appendage was normal The mitral valve measured 9 5 cm, its leaflets were thickened and the chordae tendineae were shortened and thickened but there was no evidence of acute or chronic inflammation The left ventricle was small in comparison with the right ventricle The aortic valves and aorta were normal The

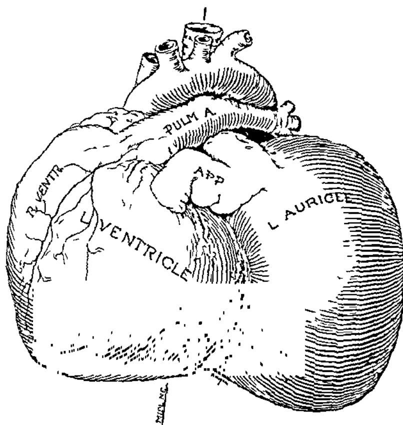


FIGURE XXX-11 Aneurysmal dilatation of the left auricle and dextrorotation of the heart

coronary arteries were smooth, but the circumflex branch of the left coronary artery was extremely small as it extended toward the saccular aneurysm and became invisible before the aneurysm was reached

Final anatomical diagnosis Dextrorotation of the heart, congenital saccular dilatation of the left auricle, thickening of the mitral and tricuspid valves, cardiac hypertrophy, and chronic passive congestion

Comment The extreme saccular dilatation of the left auricle, which was supplied by an abnormally small blood vessel, indicated that the aneurysm was probably congenital in origin The fact that the left auricular appendage appeared normal substantiated this opinion The incomplete dextrocardia was clearly a congenital abnormality This added confirmatory evidence that the abnormality of the left auricle was also congenital

The extreme dextrorotation of the heart and the abnormal position of the left auricle prevented the displacement of the esophagus and hence masked the clinical diagnosis

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CHAPTER XXXI

CORRECTED TRANSPOSITION

TRANSPOSITION OF THE VENTRICLES COMBINED WITH TRANSPOSITION OF THE GREAT VESSELS

CORRECTED transposition of the great vessels is the name given to the condition in which the aorta, although it arises from the ventricle which receives oxygenated blood, lies to the left of and anterior to the pulmonary artery, and the pulmonary artery, which arises posteriorly and to the right, receives the venous blood returned by the superior vena cava and the inferior vena cava to the right auricle Heretofore this malformation has been regarded as a rare anatomical curiosity It is, however, not as rare as was formerly believed Anderson, Lillehei, and Lester[1] have recently reported twenty-one cases in which they analyzed the clinical findings Since this report, many other cases have come to light The recognition of the condition becomes of clinical importance in the surgical correction of malformations, especially of ventricular septal defects

EMBRYOLOGY

The malformation involves an abnormal torsion of the bulboventricular loop The primitive cardiac loop swings to the left in the normal fashion but, as the bulboventricular loop forms, it is levorotated on its axis in a counterclockwise direction, so that the two ventricles lie side by side Normally the right ventricle lies anterior to the left ventricle In this malformation the ventricle which receives venous blood from the superior vena cava and inferior vena cava, and from which the pulmonary artery arises, lies to the right, and the ventricle which receives oxygenated blood returned from the lungs lies to the left It is from the latter ventricle that the aorta arises The interventricular septum lies more nearly perpendicular than parallel to the anterior chest wall Thus the abnormality primarily concerns the bulboventricular loop rather than the great vessels The rotation of the ventricular loop is such that the "left" ventricle occupies a lateral position rather than the normal posterior position

NATURE OF THE MALFORMATION

The feature of diagnostic importance is the altered position of the aorta and the pulmonary artery The aorta lies to the left and anterior to the pulmonary

artery, nevertheless, the aorta receives fully oxygenated blood from the left auri-
cle The pulmonary artery, which lies to the right and posterior to the aorta, re-
ceives the venous blood from the right auricle Although the aorta arises to the
left of the pulmonary artery, it usually arches to the left in the normal manner
The pulmonary artery also bifurcates in the normal manner Inasmuch as the
pulmonary artery lies to the right of the aorta, it is the left pulmonary artery, not
the right, which passes behind the ascending aorta as it courses to the lungs (see
Figure XXXI-1)

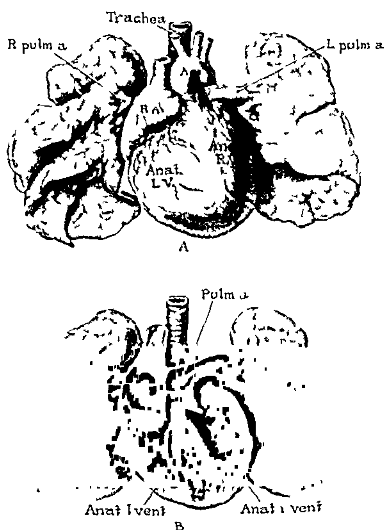


FIGURE XXXI-1 Corrected transposition combined with a ventricular septal defect and a small defect in the auricular septum (same patient as in Figure XXXI-4) Infant

The most striking of all features is that the anatomical structure of the two ventricles is transposed The smooth walled chamber with two papillary muscles is the chamber which lies to the right The wall of this chamber is composed of the deep spiral muscle bundles which normally surround the left ventricle This ventricle is, however, the one which receives the venous blood and from which the pulmonary artery arises In contrast to this, the ventricle from which the aorta arises has the anatomical structure of the right ventricle and the diffuse and extensive trabeculation characteristic of that chamber Moreover, the band of muscle known as the crista supraventricularis, which is regarded as a characteristic feature of the right ventricle, lies in this chamber at the base of the aorta This chamber appears to wrap around the central chamber as the right ventricle normally wraps around the left In addition, the mitral and tricuspid valves are transposed the mitral valve leads from the right auricle into the pulmonary ventricle and the tricuspid valve leads from the left auricle into the systemic ventricle. The coronary arteries, although they arise from the aorta, have an abnormal distribution The right coronary artery, that is, the one which arises to the right and in close proximity to the pulmonary artery, supplies both the pulmonary ventricle and the anterior portion of the systemic ventricle, whereas the left coronary artery supplies the posterior portion of the systemic ventricle and also the posterior portion of the pulmonary ventricle

COURSE OF THE CIRCULATION

The course of the circulation is not altered by the malformation The blood from the right auricle flows into the ventricle from which the pulmonary artery arises Consequently venous blood is directed to the lungs, where it is oxygenated. The oxygenated blood is returned by the pulmonary veins to the left auricle, thence it flows into the ventricle from which the aorta arises It follows that fully oxygenated blood is pumped out through the aorta to the body and the venous blood is returned in the normal manner by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again, as shown in Diagram XXXI–1

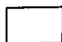
PHYSIOLOGY OF THE MALFORMATION

The basic physiology of the malformation is normal Any abnormality in the physiology is produced by an associated defect

DIAGNOSTIC PROBLEM

The diagnostic problem is twofold first, the analysis of the underlying struc

Diagram xxxi–1

 Arterial blood (fully saturated)

 Venous and arterial blood
Cyanosis visible

 Small admixture of venous blood
No visible cyanosis

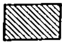
 Venous blood

DIAGRAM XXXI-1

Corrected transposition

In this malformation although the pulmonary artery arises from the anatomical left ventricle and the aorta arises from the anatomical right ventricle it is the anatomical left ventricle which receives the blood from the right auricle and the anatomical right ventricle which receives the blood from the left auricle Therefore the course of the circulation is basically normal

The blood from the right auricle flows into the anatomical left ventricle and is pumped out through the pulmonary artery to the lungs where it is oxygenated and returned in the normal fashion to the left auricle The blood in the left auricle flows into the anatomical right ventricle and is pumped out through the aorta to the systemic circulation and is returned by the superior vena cava and the inferior vena cava to the right auricle There the cycle starts again

Clinical diagnosis The condition is to be suspected in an apparently normal patient when the pulmonic second sound is markedly accentuated and the electrocardiogram shows a tendency to a left axis deviation and left ventricular hypertrophy The diagnosis is readily confirmed by angiocardiography

ture of the heart and the position of the great vessels and the ventricles, second, the nature of the associated malformation

It is, however, important to emphasize that it is usually the associated malformation which brings the patient to the doctor The abnormal rotation of the heart itself causes no symptoms Therefore, unless one is aware of and on the lookout for such an abnormality, the condition may not be detected

CLINICAL FINDINGS

The symptoms which bring the patient to the doctor depend upon the associated malformation, be it a large ventricular septal defect, pulmonary stenosis, an abnormality of either of the atrioventricular valves or even some malformation which causes persistent cyanosis

The only abnormal finding on routine physical examination is the accentuation of the second sound to the left of the sternum This may lead the doctor to suspect pulmonary hypertension

CARDIAC FINDINGS

The accentuation of the second sound at the base of the heart to the left of the sternum is the most striking clinical finding The sound is accentuated because it is produced by the closure of the aortic valve, which occupies the position of the normal pulmonary valve Indeed, the accentuation of the second sound to the left of the sternum may be quite as loud as in primary pulmonary hypertension

The closure of the pulmonary valve is both palpable and audible further to the right than is usual If there is pulmonary hypertension, the second sound over the aortic area may equal that over the pulmonary area

The thrills and murmurs produced by these two valves are similarly displaced

X RAY AND FLUOROSCOPIC FINDINGS

Although the malformation in itself does not cause cardiac enlargement, persons with this condition who seek medical attention frequently have slight to moderate cardiac enlargement Although the cardiac silhouette may appear basically normal, the contour at the base of the heart is usually abnormal (see Figure XXXI–2) The shadow immediately above the base of the heart to the left of the sternum is formed by the aorta, not by the pulmonary artery (see Figure XXXI–3) Anderson et al [1] have emphasized that this shadow may have a convex curve, a concave curve, or even a straight margin, depending on the size of the ventricle and the exact course of the aorta

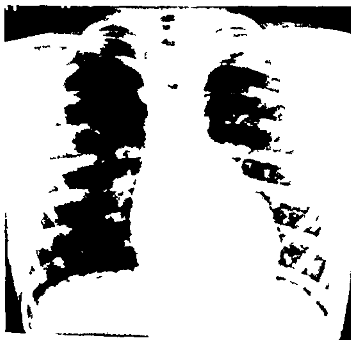


FIGURE XXXI-2. Corrected transposition (same patient as in Figure XXXI-6) Child

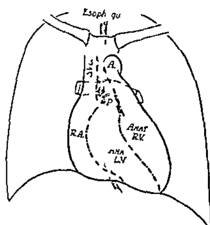


FIGURE XXXI-3 Corrected transposition Child

The abnormal position of the pulmonary artery offers a clue to the diagnosis Even though the pulmonary artery is enlarged, there is no corresponding fullness of the pulmonary conus Indeed, the vascular markings may appear increased and yet the shadow at the base of the heart may appear abnormally narrow, as in a transposition of the great vessels (see Figure XXXI-4) Similarly in cases of pulmonary stenosis, the poststenotic dilatation of the pulmonary artery is not visible and yet the branches of the pulmonary artery appear to be normal or increased in size

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiographic findings may also be of diagnostic aid Anderson et al [1] have reported the frequent occurrence of prolongation of the P-R interval combined with high P waves There may be only a first degree heart block or there may be a second degree block or even a complete dissociation Furthermore, a patient with a normal conduction time may, over a period of years, develop a partial heart block which may progress to a complete heart block

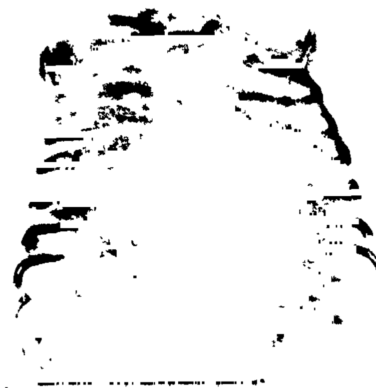


FIGURE XXXI-4 Corrected transposition combined with a ventricular septal defect and a small defect in the auricular septum (same patient as in Figure XXXI-1) Infant

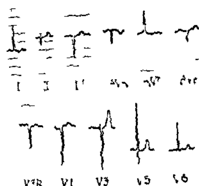


FIGURE XXXI-5 Corrected transposition

Lead I and AVL show a normal RS *pattern*, whereas Lead III and AVR and AVF show the QR pattern so frequently seen in left ventricular dominance[1] The unipolar precordial leads may offer a clue to the diagnosis when there is a QR *pattern in* V_1 *combined* with an RS pattern in V_6 and the T waves are upright across the entire precordium from V_3 on the right through V_6 *on* the left (see Figure XXXI-5)

It should be borne in mind that the functional right *ventricle has the anatomical structure* of the left ventricle and that the anatomical right ventricle, which is the functional left ventricle, occupies a position in the chest similar *to* that of the normal left ventricle (see Figure XXXI-3) Nevertheless, when the pulmonary ventricle (anatomical left ventricle) undergoes hypertrophy, it is apparently reflected in the electrocardiogram by the pattern which is normally associated with that of left ventricular hypertrophy Indeed, the author has seen one instance in which V_6 showed the pattern of so-called left ventricular strain with a tall R, a wide RS *complex and slight inversion* of the T wave In this in*stance the wall of the pulmonary ventricle, which was the anatomical left* ventricle but the functional right ventricle, was of *greater* thickness than the wall of the systemic ventricle This suggests that the spiral muscle bundles of the anatomical left ventricle give the *pattern of left ventricular preponderance* in the electrocardiogram Certain it is that in this malformation evidence of so-called left ventricular strain should not be interpreted as *evidence of* hypertrophy of the systemic ventricle Furthermore electrocardiographic evidence of greater left than right ventricular dominance in a patient with *marked* accentuation of the second sound to the left of the sternum should arouse suspicion of the existence of a corrected transposition

SPECIAL TESTS

Angiocardiography is the easiest method by which to demonstrate the altered position of the great vessels An angiocardiogram taken in the anterior-posterior position usually shows that the pulmonary artery occupies a more medial position than is normal, furthermore, when the aorta is visualized, it is seen to lie to the left of the pulmonary artery and forms the upper margin of the cardiac silhouette to the left of the sternum The diagnosis is confirmed by the abnormal contour of the right ventricle In place of the normal trabeculation, the outline of the right ventricle is smooth and has a tail like configuration" as emphasized by Anderson et al [1] *(see Figure xxvi-6)*

Cardiac catheterization may be of aid in the demonstration of the abnormal position of the pulmonary artery Indeed, with the usual swing of the catheter along the outer margin of the ventricle, the catheter fails to enter the pulmonary artery Inability to enter the pulmonary artery in its normal position should arouse suspicion of a corrected transposition, especially if there is no clear evidence of pulmonary stenosis or of markedly reduced pulmonary blood flow Further exploration with the catheter may reveal the medial position of the pulmonary orifice *Occasionally the pulmonary artery is entered so easily that its* abnormal position is not detected

DIAGNOSIS

The marked accentuation of the second sound to the left of the sternum, when combined with absence of electrocardiographic evidence of right ventricular hypertrophy, should suggest the diagnosis of an abnormal rotation of the great vessels and of the ventricles Simple awareness of the possibility aids in diagnosis

Although the contour of the heart varies with the nature of the concomitant malformation, difficulty in the visualization of the main pulmonary artery to the left of the sternum, in a non-cyanotic patient who has evidence *of a left-to-*right shunt, should arouse suspicion

The electrocardiogram frequently aids in the diagnosis In an uncomplicated corrected transposition of the great vessels there is evidence of left ventricular dominance When this occurs in an asymptomatic patient with clinical evidence of primary pulmonary hypertension, it should suggest that the position of the great vessels is transposed The occurrence of prolongation of the P-R interval, combined with high P waves in V_1 or evidence of second or third degree heart block, is also suggestive of this anomaly Generally both the stand



Dextrogram



Levogram

FIGURE XXXI-6 Corrected transposition (same patient as in Figure XXXI-[illegible]) Child

ard leads and the unipolar precordial leads fail to conform to the expected pattern

The diagnosis is most readily confirmed by angiocardiography, which shows the abnormal positions of the pulmonary artery and the aorta, and the tail like configuration of the "right" ventricle

DIFFERENTIAL DIAGNOSIS

Primary pulmonary hypertension is frequently confused with this malformation because of the accentuation of the second sound at the base of the heart to the left of the sternum The electrocardiogram in a corrected transposition usually shows evidence of left ventricular dominance, whereas in primary pulmonary hypertension there is evidence of right ventricular strain

Pulmonary stenosis combined with a corrected transposition of the great vessels may be confused with primary pulmonary hypertension if at catheterization the pulmonary artery is not entered, because in both conditions the pressure in the right ventricle is elevated The electrocardiogram usually offers a clue to the diagnosis In a corrected transposition combined with pulmonary stenosis, the electrocardiogram seldom shows the pattern of right ventricular hypertrophy

Complete transposition of the great vessels may occasionally be confused with a corrected transposition because of the absence of fullness of the pulmonary conus combined with evidence of increased hilar shadows The absence of cyanosis sharply differentiates a corrected transposition of the great vessels from the usual complete transposition of the great vessels

COMMONLY ASSOCIATED MALFORMATIONS

Inasmuch as the malformation represents a basic abnormality in the rotation of the primitive bulboventricular loop, it is not surprising to find that the condition is frequently associated with additional abnormalities Almost any type of abnormality may occur with it The two most common concomitant malformations are defects in the ventricular septum and pulmonary stenosis, malformations of the mitral and tricuspid valves are also common Furthermore, in a corrected transposition of the great vessels, Ebstein's anomaly of the tricuspid valve may occur in the systemic ventricle,[2] which is the anatomical right ventricle (see Figure XXXI-7) Each of the various associated malformations affects the course of the circulation in its specific manner

TREATMENT

The condition itself requires no treatment Nevertheless, recognition of the

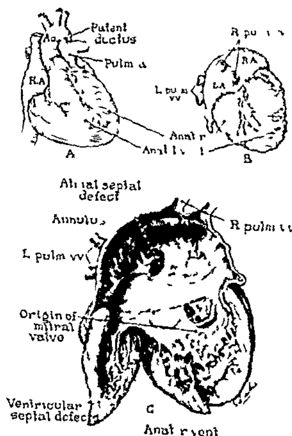


FIGURE XXXI–7 Corrected transposition (*transposition of the ventricles*) and Ebstein's anomaly of the tricuspid valve in the functional left ventricle The great vessels are again transposed the aorta *arises from* the anatomical left ventricle which receives blood *from* the right auricle the pulmonary artery *arises from* the anatomical right ventricle which receives blood from the pulmonary veins

Note Dr Jesse Edwards studied this specimen and agreed with the above analysis

condition is of tremendous importance if the concomitant malformation requires surgical correction The anomalous origin and the distribution of the coronary arteries are of great concern when a ventriculotomy is to be performed Furthermore, incision of spiral muscle bundles of the left ventricle is less well tolerated than is the incision of the musculature of the normal right ventricle Consequently, although it is usually preferable to incise the right ventricle rather than the left ventricle for the closure of a ventricular septal defect, in this malformation incision of the systemic ventricle may be preferable, as it has the anatomical structure of the right ventricle Nevertheless, the abnormal course of the right coronary artery and its main branches poses a difficulty For this reason, Lillehei[1] recommends an auricular approach for closure of ventricular septal defects in patients with this type of corrected transposition

SUMMARY

Corrected transposition of the great vessels is the name given to the condition in which a transposition of the great vessels is combined with a transposition of the ventricles so that, although the aorta arises in the position of the normal pulmonary artery from a ventricle which has the internal structure of the right ventricle, it still receives oxygenated blood from the left auricle and directs the blood to the systemic circulation The pulmonary artery lies to the left of the aorta and arises from the ventricle which has the anatomical structure of the left ventricle, nevertheless, this ventricle receives venous blood from the right auricle and directs it to the lungs The deep spiral muscle bundles surround the venous ventricle and the crista supraventricularis lies in the systemic ventricle The mitral and tricuspid valves are also transposed The coronary arteries follow an abnormal course It is the right coronary artery which has two main branches one supplies the pulmonary ventricle and the other supplies the anterior surface of the systemic ventricle

The malformation in itself causes no difficulty but it is frequently associated with some other malformation of the heart The course of the circulation is unaltered by this anomaly

It is usually the concomitant malformation which brings the patient to the doctor

The outstanding finding of clinical importance is the accentuated second sound to the left of the sternum in the absence of electrocardiographic evidence of 'right' ventricular hypertrophy

The diagnosis should be suspected when the second sound over the pulmo-

nary area is markedly accentuated and the electrocardiogram shows evidence of left rather than right ventricular dominance

The diagnosis is readily established by angiocardiography, which shows that the pulmonary artery occupies a medial position and the aorta lies to the left of the pulmonary artery

The malformation requires differentiation from primary pulmonary hyper tension, from pulmonary stenosis, and from a complete transposition of the great vessels.

The condition itself causes no difficulty but becomes of clinical importance in the correction of associated malformations

References

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CHAPTER XXXII

ANOMALIES OF THE SYSTEMIC VENOUS RETURN

THE major anomalies of the systemic venous return which are of importance in relation to cardiac anomalies are those which concern the venae cavae To understand these abnormalities a knowledge of the embryology of the venae cavae is essential Section A discusses the anomalies of the superior vena cava and the inferior vena cava, Section B the absence of the inferior vena cava

EMBRYOLOGY*

The outstanding characteristic of the systemic venous pattern of the young embryo, according to Patten, is its bilateral symmetry In the initial stages the *paired cardinal veins* from the anterior and posterior parts of the embryo become confluent and enter the medially placed sinus region of the primitive tubular heart (see Figure XXXII-1 A) In contrast to this, in the fully developed fetus, only the peripheral channels remain paired and the main venous channels which return the blood to the heart are single vessels, namely, the superior vena cava and the inferior vena cava, these are normally located on the right side of the body and empty into the right auricle (see Figure XXXII-1 F)

The formation of the superior vena cava is relatively simple The main tributaries from the anterior part of the body are the external and internal jugular veins and the subclavian veins *The internal jugular veins* are the anterior portions of the original anterior cardinal veins The shift of the venous blood flow to enter the right side of the heart is accomplished by the formation of a new vessel between the right and left anterior cardinal veins, namely, the *left innominate vein* as this vessel forms, the lower portion of the left anterior cardinal vein degenerates The right anterior cardinal vein between the union of the jugular veins and the right subclavian vein becomes the *right innominate vein* (see Figure XXXII-1 F) The vessel which extends from the point of union of the two innominate veins to the heart is the *superior vena cava*

The formation of the inferior vena cava is far more complex, as it develops

*This entire section is derived from Patten s *Human Embryology* (pp 638-644) for a more detailed discussion the reader is referred to this source[1] and to other authorities on embryology

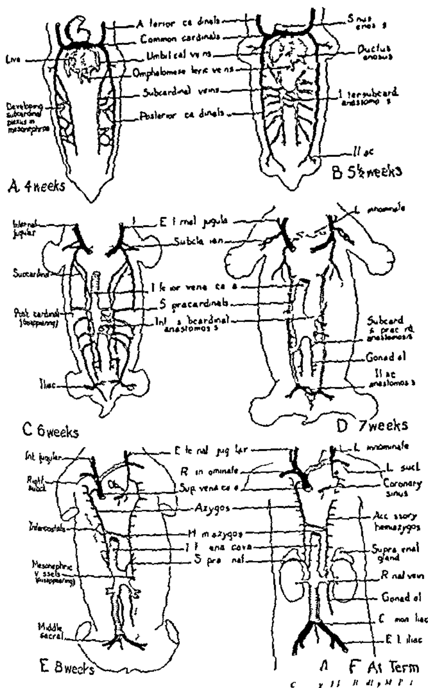


Figure xxxii-1 Embryological development of the inferior vena cava

in three separate parts the mesenteric portion, the hepatic portion, and the postrenal portion

In the tiny embryo the *posterior cardinal veins* bring the blood back to the heart from the posterior part of the body The posterior cardinal veins develop in association with the mesonephroi As the latter degenerate, the posterior cardinal veins tend to degenerate and the *subcardinal veins* and the *supracardinal veins* enlarge Broadly speaking, the *subcardinal veins* and their tributaries return the blood from the abdominal organs to the heart and the *posterior portions of the supracardinal veins* drain the pelvis and the lower extremities The *anterior segments of the supracardinal veins* develop into the azygos veins (Figure XXXII–I E and F)

The subcardinal veins initiate the diversion of blood from the posterior cardinal veins As the posterior cardinal veins degenerate, the subcardinal veins enlarge Originally the subcardinal veins form as an irregular plexus of small veins which develop numerous anastomoses with the posterior cardinal vein As the subcardinal veins enlarge, many of the small plexuses become confluent and intracardinal anastomoses develop These channels, together with the subcardinal veins, form the *subcardinal sinus* This sinus, which lies at the level of the future renal vessels, drains medially rather than laterally The posterior cardinal veins in this region disappear

The subcardinal sinus, as it comes to carry an increased volume of blood, finds a new pathway to the heart The plexus on the right side comes to lie close to the liver and it eventually connects with the numerous small plexuses which are simultaneously developing in the liver Once the subcardinal sinus connects with these vessels in the liver, the new channel grows rapidly and forms the *mesenteric part of the inferior vena cava*

Within the liver the stream of blood from the mesenteric part of the inferior vena cava at first follows devious channels as it returns the blood to the sinus venosus As the volume of blood increases, a new channel is formed within the liver, this new pathway forms the *hepatic part of the inferior vena cava*

A third set of veins enters into the formation of the distal part of the inferior vena cava and also into the azygos system, namely, the *supracardinal veins*

The posterior portions of the supracardinal veins that is, the portions of the supracardinal veins which lie distal to the subcardinal sinus, become the principal pathways by which blood is returned from the pelvis and the lower extremities With the anastomosis between the right and left iliac veins, an increasing volume of blood is returned by the right subcardinal vein, which becomes enlarged to form the *postrenal portion of the inferior vena cava*

Figure XXXII-1 illustrates the various stages in the development of the superior vena cava and the inferior vena cava Diagrams A and B show the anterior and posterior cardinal veins, Diagrams C and D show the development of the superior vena cava and also the disappearance of the posterior cardinal veins and the formation of the mesenteric portion of the inferior vena cava In Diagram E the superior vena cava has formed, the azygos system is developing, and so is the postrenal portion of the inferior vena cava Diagram F shows the venous pathways which are normally present at term

The azygos veins also return some blood from the abdomen to the right auricle (see Figure XXXII-1 E and F) The azygos veins develop from the *anterior portions of the supracardinal veins*, which lie cephalic to the level of the subcardinal sinus The point of entrance of the azygos vein into the superior vena cava indicates the original point of transition from the anterior cardinal veins to the common cardinal vein

A *Anomalies of the Superior Vena Cava and the Inferior Vena Cava*

A persistent left superior vena cava is relatively common The left superior vena cava may enter the right auricle by way of the coronary sinus or it may open directly into the left auricle

The anomalous entrance of the left superior vena cava into the coronary sinus causes no difficulty The condition is to be suspected when a shadow extends downward from the inner margin of the left clavicle to the upper margin of the cardiac silhouette The condition is readily detected by cardiac catheterization or angiocardiography when these tests are performed through the left brachial or radial vein, as demonstrated in Figure XXXII-2 Inasmuch as it is physiologically inconsequential whether the left superior vena cava drains into the innominate vein, and hence into the right auricle by way of the superior vena cava, or directly into the right auricle through the coronary sinus, the condition calls for no treatment The entrance of the left superior vena cava into the coronary sinus is, however, of clinical importance in the correction of malformations in which an extracorporeal circulation is used, because this anomaly greatly increases the volume of blood returned to the heart through the coronary sinus

An anomalous drainage of the left superior vena cava into the left auricle causes some venous blood to be directed to the systemic circulation This anomaly occurs most frequently with isolated dextrorotation of the heart and with a situs inversus and levocardia (see Chapters XXXIII and XXXIV) The condition

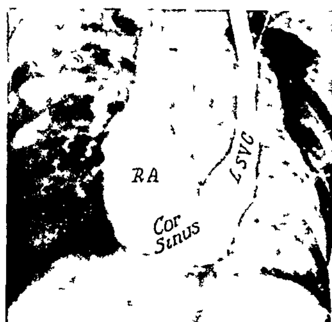




FIGURE XXXII-2 Anomalous drainage of the left superior vena cava into the coronary sinus Child

may be detected by cardiac catheterization and is readily seen in an angiocardiogram if the dye is injected through a vein in the left arm (see Figure xxxii-3)

In anomalous drainage of the superior vena cava and the inferior vena cava into the left auricle is a rare condition When this does occur, the right auricle lacks its normal inflow tracts, under such circumstances the only way for blood [illegible]

[illegible]

the right ventricle is usually underdeveloped There may be tricuspid atresia

The author has seen this type of anomaly only twice In one instance the tricuspid valve was normally formed and opened into an abnormally small right ventricle and in addition the pulmonary artery was atretic at its base, the aorta over rode the ventricular septum and received the blood from both ventricles The course of the circulation is shown in Diagram xxxii-1 The clinical and cardiac findings were similar to those of defective development of the right ventri

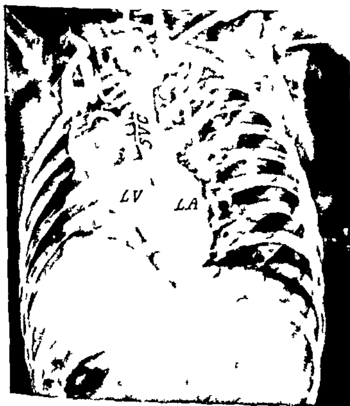


Figure xxxii-3 Anomalous drainage of the left superior vena cava into the left auricle Child

Diagram xxxii–1

Head and upper extremities

Ductus Arteriosus

R Lung

P.A.

S.V.C.

Aorta

Pul. A.

L Lung

PV

Pul V

L.A.

Descending Aorta

R.A.

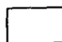
L.V.

R.V.

I.V.C.

Remnant of Pulmonary Artery

Trunk and lower extremities

 Arterial blood (fully saturated)

 Venous and arterial blood
Cyanosis visible

 Small admixture of venous blood
No visible cyanosis

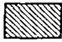
 Venous blood

Diagram XXXII–1

Anomalous drainage of the superior and inferior venae cavae into the left auricle combined with an underdeveloped right auricle and right ventricle dextroposition of the aorta a high ventricular septal defect and pulmonary atresia

In this malformation the superior and inferior venae cavae drain into the left auricle. The only way for the blood to reach the right auricle is through a defect in the auricular septum The malformation usually occurs in association with some serious malformation of the right ventricle, such as pulmonary atresia and a dextroposition of the aorta, or a non functioning right ventricle.

All the blood from the superior vena cava and the inferior vena cava flows directly into the left auricle and some blood from the left auricle flows into the right auricle through the defect in the auricular septum The blood in the right auricle flows into the right ventricle. Inasmuch as there is pulmonary atresia, the only way for the blood to escape is through the dextroposed aorta Here it meets the blood from the left ventricle and flows out through the aorta to the systemic circulation Because of the pulmonary atresia the pressure in the pulmonary artery is low and some blood will flow from the aorta through the ductus arteriosus to the pulmonary artery and thence to the lungs, where it is oxygenated All the blood which reaches the lungs is returned in the normal fashion to the left auricle The blood which flows into the left ventricle is pumped out through the aorta to the systemic circulation and is returned by the superior and inferior venae cavae. These vessels however empty into the left auricle. Thus all the blood from the systemic circulation, as well as all the blood from the lungs, is returned to the left auricle The only way for blood to reach the right auricle is by way of the left auricle. As mentioned above part of the blood from the left auricle flows into the right auricle and thence to the right ventricle the remainder of the blood in the left auricle flows into the left ventricle and is pumped out into the systemic circulation.

There is complete admixture of arterial and venous blood in the left auricle Furthermore, less blood reaches the lungs for oxygenation than reaches the systemic circulation hence a small volume of oxygenated blood is mixed with a large volume of venous blood Cyanosis is intense.

Clinical diagnosis The structure of the ventricles varies with the associated malformation The condition places a severe strain upon the circulation it is compatible with life for only a few months

cle Angiocardiography would have shown that the left auricle was visualized before the right auricle This might have offered a clue to the diagnosis

Since such an infant obviously suffers from reduced blood flow, a systemic pulmonary anastomosis would improve the circulation to the lungs, but the heart is so grossly defective that the malformation is often not compatible with life for more than a few months

In the other instance which the author has seen, the patient also suffered from decreased pulmonary blood flow and had a Blalock-Taussig anastomosis in 1950 Although his heart is greatly enlarged, he is still living in 1960, there fore the exact nature of his malformation is not known

An anomalous drainage of the inferior vena cava into the left auricle only rarely occurs as an isolated anomaly Gardner and Cole[2] have reported one such case the woman had been cyanotic throughout her life and had suffered from slight shortness of breath She had married at eighteen years of age, had had numerous miscarriages, and one living child She died suddenly at thirty two years Autopsy showed a healed myocardial infarction The sole anomaly in the heart was the entrance of the inferior vena cava into the left auricle This case clearly demonstrates that the possibility of the anomalous entrance of the inferior vena cava into the left auricle should be considered in a cyanotic patient in whom no obvious cause of the venous arterial shunt can be found

An anomalous drainage of the inferior vena cava into the left auricle or the left side of the common auricle may occur with dextrorotation of the heart or with a situs inversus and levocardia (see Chapters XXXIII and XXXIV)

Minor anomalies of the inferior vena cava are common[3] but are unimportant in relation to cardiac malformations The only other major anomaly of real im portance is absence of the inferior vena cava

SUMMARY

The anomalies of the systemic venous return which are of importance in re lation to cardiac abnormalities are those which concern the superior vena cava and the inferior vena cava

A brief review of the embryology of these vessels shows that the formation of the superior vena cava is simple and that of the inferior vena cava is complex The latter forms in three main sections the hepatic portion, the mesenteric por tion, and the postrenal portion The anomalies associated with the inferior vena cava are correspondingly more complex than are those of the superior vena cava

The major anomaly of the superior vena cava is persistence of the left su perior vena cava, which may enter into the coronary sinus or into the left auricle

... instances open directly into the left auri ...

... oxygen unsaturation of the arterial blood in whom no other ... arterial shunt can be demonstrated

B Absence of the Inferior Vena Cava

Absence of the inferior vena cava is a rare anomaly When the mesenteric portion of the inferior vena cava fails to connect with the liver, the hepatic veins drain the blood from the liver into the lower part of the right auricle They may enter as several small vessels or they may become confluent and enter as a single large vessel The remainder of the blood from the abdomen and the lower extremities is returned by the azygos veins which always enlarge when the inferior vena cava is absent The azygos vein usually opens into the superior vena cava in the normal manner but its opening may become incorporated into the upper portion of the right, or "caval," auricle The latter situation is relatively common where there is dextrorotation of the heart

NATURE OF THE MALFORMATION

Absence of the inferior vena cava occurs most frequently in dextrorotation of the heart and in a situs inversus and levocardia, in both conditions the relation of the auricles to the abdominal viscera is abnormal When there is dextrorotation without a situs inversus, the right auricle lies to the right of the sternum and the liver occupies its normal position on the left, whereas the reverse is true with a situs inversus and levocardia, the abdominal organs have been reversed and the heart has rotated back to the left Thus in both instances the right auricle no longer lies above the liver In order for the inferior vena cava to develop normally it is essential for the right auricle to lie on the same side of the body as the liver In neither of the above anomalies is this true, consequently in neither of these anomalies can the inferior vena cava enter the right auricle in the nor...

... with its hepatic portion Under such circumstances there is no inferior vena cava Absence of the inferior vena cava may occasionally occur as an isolated anomaly when the abdominal viscera are normally placed

The azygos vein always enlarges when the inferior vena cava is absent, as it serves as the main pathway by which the blood is returned to the heart from the abdominal organs and from the pelvis and the lower extremities The azygos vein usually opens into the superior vena cava in its normal position

The blood from the liver is, however, returned to the heart by the hepatic veins, which pierce the diaphragm and enter the auricle which lies adjacent to it. Thus, if there is a partial situs inversus of the abdominal organs and the liver occupies a mid line position, the hepatic vein may develop normally on the left and enter the right auricle in the normal fashion even though the inferior vena cava is absent. If the liver lies on the left, the hepatic veins pierce the diaphragm on the left and enter the auricle adjacent to it. If the auricles are not transposed, the hepatic vein may enter the "left" auricle close to the entrance of the pulmonary veins, as shown in Figure xxxiii-5 (see page 972)

CLINICAL FINDINGS

Absence of the inferior vena cava, in itself, causes no symptoms. The possibility, however, should be considered whenever the relation of the heart to the abdominal viscera is abnormal or when the x ray contour is abnormal.

The condition becomes of clinical importance in the correction of malformations with the use of a pump and oxygenator. Not only is the main drainage from the lower part of the body returned high up into the right auricle but, in addition, the hepatic veins return a considerable volume of blood to that chamber. The hepatic veins may enter the right auricle through a single orifice or through multiple orifices which open into the base of the right auricle in the location usual for the inferior vena cava.

CARDIAC FINDINGS

A gross malformation of the heart is likely to overshadow evidence of an abnormality of the inferior vena cava. The possibility of absence of the inferior vena cava should be suspected in a dextrorotation of the heart and in a situs inversus with levocardia (see Chapters xxxiii and xxxiv)

X RAY AND FLUOROSCOPIC FINDINGS

The characteristic x ray findings were first reported by Downing[4]. In the anterior posterior view a slight bulge is visible in the superior vena cava just above the right auricle (see Figure xxxii-4)

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram reflects the other abnormalities of the heart but is not affected by the anomalous course of either of the venae cavae



FIGURE XXXII–4 Absence of the inferior vena cava Child

Note anomalous drainage of a persistent right posterior cardinal vein into the right auricle near the superior vena cava

SPECIAL TESTS

Cardiac catheterization offers a clue to the diagnosis The absence of the inferior vena cava is readily demonstrated if the catheterization is performed through the saphenous vein In Downing's[4] first case, he passed a catheter through the left superior vena cava, which opened into the coronary sinus, and up through the right auricle and down the azygos vein He confirmed the absence of the inferior vena cava by angiocardiography If a catheter is passed into the azygos vein directly from the right auricle, the demonstration of the abnormal position of the azygos vein should arouse suspicion that the inferior vena cava may be absent.

Angiocardiography performed through the saphenous vein also readily reveals the existence of this anomaly[4] Figure xxxii–5 demonstrates the absence of the inferior vena cava in a patient in whom a selective angiocardiography was performed through the saphenous vein

DIAGNOSIS

The clinical diagnosis is not easy When the heart occupies its normal posi

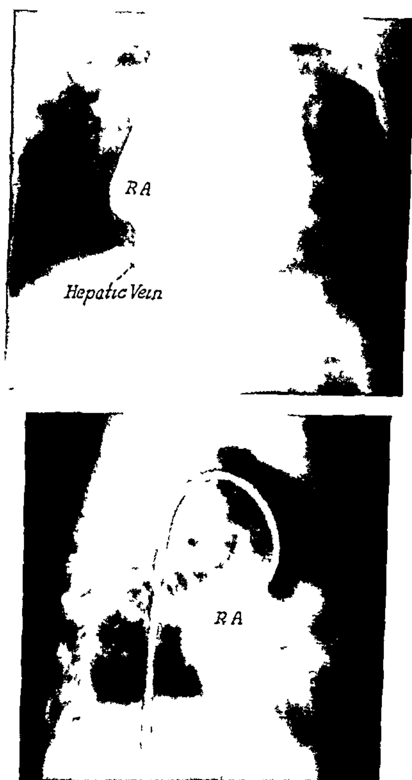


FIGURE XXXII–5 Absence of the inferior vena cava Child
Note the course taken by the catheter

tion, the absence of the inferior vena cava may be suggested by the abnormal contour of the right auricle The possibility of this anomaly should always be borne in mind when there is a dextrorotation of the heart or a situs inversus with levocardia.

DIFFERENTIAL DIAGNOSIS

The condition is more frequently overlooked than it is confused with other malformations

A total anomaly of the pulmonary venous return to the superior vena cava may cause a similar dilatation of the superior vena cava in the x ray The two conditions are, however, seldom confused because a total anomaly of the pulmonary venous return is always associated with increased pulmonary blood flow and electrocardiographic evidence of right ventricular hypertrophy

COMMONLY ASSOCIATED ANOMALIES

It is the associated anomaly which brings the patient to the doctor In most cases the absence of the inferior vena cava is merely an incidental finding It is the underlying malformation which causes the symptoms and which requires treatment

TREATMENT

There is none and none is needed Nevertheless, knowledge of the existence of the abnormality may be of great importance in cardiac surgery The azygos vein, when it enters normally into the superior vena cava, can usually be ligated with impunity If, however, the azygos vein is abnormally large or opens into the right auricle, it is in all probability the principal pathway by which the blood from the trunk and the lower extremities is returned to the heart Under such circumstances the azygos vein should never be ligated unless the surgeon is certain that the inferior vena cava is normal

PROGNOSIS

The prognosis varies with the associated cardiac anomaly

SUMMARY

The major anomaly of the inferior vena cava is its absence When this occurs as an isolated anomaly, the azygos vein usually opens normally into the superior vena cava When the condition occurs in association with dextrorotation of the

heart or a situs inversus with levocardia, the azygos vein may open into the superior vena cava or directly into the right auricle In both instances, the mesenteric portion of the inferior vena cava is absent but the hepatic veins enter the right auricle in the normal manner

Absence of the inferior vena cava may be suspected when an x ray of the heart reveals an abnormal bulge in the superior vena cava just above the right auricle

The abnormality can be proven by cardiac catheterization or by angiocardiography, provided the test is performed through the saphenous vein

The condition occasionally requires differentiation from a total anomaly of the pulmonary venous return to the superior vena cava

In itself the condition causes no symptoms and requires no treatment but the diagnosis is important in patients who require cardiac surgery If the azygos vein is seen to enter the right auricle, it should never be ligated, as it is probably the main pathway for the return of blood from the trunk and the lower extremities Indeed, if the azygos vein is abnormally large, the possibility of absence of the inferior vena cava should always be excluded before the azygos vein is ligated

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CHAPTER XXXIII

DEXTROCARDIA AND DEXTROROTATION

A TRUE dextrocardia occurs when there is a complete reversal of all the organs of the body The condition is known as situs inversus Under such circumstances the position of the heart and the abdominal organs are the mirror image of the normal In contrast to this, dextrorotation of the heart means that the heart has twisted abnormally to the right Furthermore, it is generally believed that, whereas complete reversal of all the organs of the body is only rarely associated with an additional abnormality of the heart, rotation of the heart alone usually results in some extremely severe cardiac malformation The latter is certainly true In the author s experience the incidence of severe malformations of the heart in cases of situs inversus is relatively high When it is recalled that a reversal in the direction of the primitive cardiac loop is an abnormality, it is not surprising that the abnormal rotation is frequently associated with additional cardiac abnormalities

When a dextrocardia occurs as the sole cardiac abnormality in combination with complete situs inversus, the diagnosis is relatively simple When, however, in addition to the dextrocardia, there is evidence of a further cardiac malformation, the condition presents one of the most complicated of diagnostic problems

A *Dextrocardia with Situs Inversus*

NATURE OF THE MALFORMATION

In a dextrocardia the primitive cardiac loop swings in the reverse *direction* to the *normal* This condition occurs with least disturbance to the embryo when *there* is a complete reversal in the position of all the organs of the body In a situs inversus the position of all the organs is the mirror image of the *normal* The heart lies on the right side instead of on the left and *the apex points to the right* The lobes of the lungs are reversed there are three lobes on the *left side* and two on the right The liver lies on the left side of the abdomen, *the spleen* on the right The gastrointestinal tract is also reversed the ascending colon lies on the left the transverse colon crosses from left *to right*, and the descending colon is on the right.

The reversal is from left to right, not from front *to back* The apex of the heart points to the right but the right *ventricle* lies in front of the left ventricle

The right auricle lies to the left of the sternum, the left auricle lies posteriorly If the auricles are enlarged, it is still the left auricle which causes the backward displacement of the esophagus The aorta arches over the pulmonary artery and descends upon the right side of the body

The only change which occurs in the structure of the heart is of purely academic interest It concerns the direction of the superficial muscle bundles It is a curious fact that the direction of the superficial muscular layer of the heart is not reversed [1] The superficial bundles run from base to apex in a clockwise direction The direction of the deep muscle bundles, however, is the mirror image of the normal

COURSE OF THE CIRCULATION

The course of the circulation is the mirror image of the normal The reversal of the position of all the organs places no abnormal strain on the heart or vascular system

PHYSIOLOGY OF THE MALFORMATION

The condition in no way affects the physiology of the circulation It is entirely normal

CLINICAL FINDINGS

The condition, when not associated with other malformations, is asymptomatic It does, however, apparently carry an increased liability to bronchiectasis [2] Furthermore, it is important for the physician to remember that, with the reversal in the position of the organs, the appendix lies in the lower left quadrant

CARDIAC FINDINGS

The condition is a test of the physician's acuity in physical diagnosis The functional capacity of the heart is unaltered There are no murmurs and no thrills Clubbing and cyanosis are absent Indeed, there may be nothing to attract attention to the dextrocardia

If the apex beat is visible, inspection will reveal the condition The point of maximal impulse is seen in the fourth or fifth interspace, close to the mid-clavicular line on the right side Palpation is also of great help in that the apical impulse is felt on the right side It is the position of the apex thrust which most often gives the clue to the diagnosis

When the physician is aware of a dextrocardia he usually experiences no

difficulty in the percussion of the heart upon the right but, when caught unawares, many a person has outlined the area of cardiac dullness *on the left* In an infant, accurate percussion of a heart of normal size is extremely *difficult* Palpation and auscultation are of far greater aid When the heart lies mainly in the right thorax, the heart sounds are better heard to the right of the sternum than to the left Therefore, whenever the heart sounds are of maximum intensity in this location, even though the other physical findings *appear to* be normal, the possibility of a dextrocardia should always be considered

Examination of the abdomen is the only clinical way to differentiate a complete situs inversus from an extreme dextrorotation or dextroversion In a situs inversus the liver lies on the left and the stomach and spleen on the right, whereas if the heart alone is rotated the positions of the abdominal viscera are normal

X RAY AND FLUOROSCOPIC FINDINGS

The demonstration by x ray that the heart and the stomach lie on the right and the liver on the left establishes the diagnosis (see Figure XXXIII-1)

A situs inversus is such a perfect mirror image of the normal that, unless careful regard is paid to the markings on the x ray film, it is easy to reverse the film and read it as heart normal in size and position

Fluoroscopic examination reveals the condition at a glance As soon as one perceives that the position of the heart is the mirror image of the normal, one should immediately try to observe the position of the liver and the stomach A barium swallow quickly reveals the position of the stomach Therefore, after the administration of barium, it is wise to form the habit of lowering the fluoroscopic screen and glancing at the stomach This is especially important in the diagnosis of a situs inversus and a levocardia (see Chapter XXXIV)

When there is a dextrocardia it is important to remember that a right aortic arch is normal Inasmuch as the superior vena cava lies on the left, the esophagus will occupy its normal mid line position, it will, however, be indented on its right margin by the aorta Furthermore, the right and left anterior-oblique positions replace each other The contour of the heart seen in the left anterior oblique position is characteristic of that normally seen in the right anterior oblique position and vice versa When there is a left aortic arch, the analysis of the relation of the heart and the aorta must be made with care because the position of the ventricles is the mirror image of the normal, whereas the apparently normal left aortic arch is in reality the equivalent of a right aortic arch

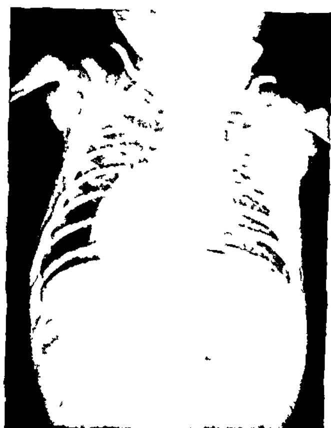


FIGURE XXXIII-1 Dextrocardia and situs inversus Infant

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram is pathognomonic A typical example of such an electrocardiogram is shown in Figure XXXIII-2 Lead I is the mirror image of the normal Leads II and III replace each other In Lead I both the P waves and the T waves are inverted and the principal deflection of the QRS complex is the mirror image of the normal The findings in Lead III are those usually seen in Lead II and the form of the deflection in Lead II is similar to that normally seen in Lead III, consequently it is Lead II, not Lead III, in which variations in the form of the complexes frequently occur with respiration No other condition produces all the above mentioned changes, an identical electrocardiographic picture is, however, produced by the incorrect application of the arm electrodes

The unipolar precordial leads should be taken over the right chest When so taken the normal V_2 on the left is the equivalent of V_1 on the right, V_3R is equivalent to the normal V_3L, and V_5R and V_6R have the same significance as V_5 and V_6 have for a patient with a normally placed heart V_3L clarifies the findings of V L, just as V_3R normally clarifies the findings of V_1 The electrocardio-

gram in Figure XXXIII–2 shows a balanced axis and evidence of "combined" hypertrophy in a patient with a dextrocardia

In order to evaluate the axis deviation the fact that the direction of the deflections in Lead I is the mirror image of the normal must be taken into account For example, in a left axis deviation the primary deflection in Lead I is downward and the S wave in Lead II is greater than one half of the S wave in Lead I Evidence of left ventricular dominance is indicated by a deep S wave in V and a tall R wave in V_5R and V_6R Figure XXXIII–3 shows such an electrocardiogram, although unfortunately V was not taken, V_3L, however, clearly shows greater left than right ventricular dominance This finding is in striking contrast to the right ventricular hypertrophy seen in V L and V_3L in Figure XXXIII–4

In contrast to these findings, when there is a right axis deviation, the R wave is greater than the S wave in Lead I and the R wave in Lead II is also greater than the S wave in that lead When there is right ventricular hypertrophy, V_2 is the lead in which the tall R wave is significant V_3L clarifies the evidence of

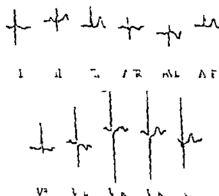


FIGURE XXXIII–2 Dextrocardia with combined hypertrophy

All electrocardiograms of patients with dextrocardia or dextrorotation are arranged so that, as the precordial leads are read from left to right the findings are first related to the anterior chamber (V_1 and V) and then to the posterior chamber (V_5R and V_6R) This order is used because it is the customary one in which the precordial leads are read

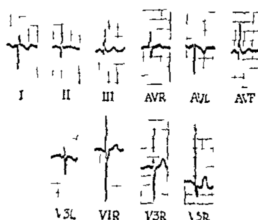


FIGURE XXXIII-3 Dextrocardia with dominance of the posterior chamber (unfortunately V2L was not taken)

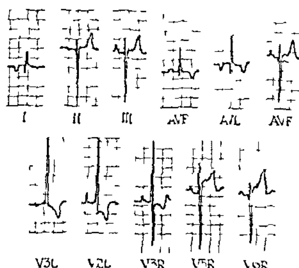


FIGURE XXXIII-4 Dextrocardia with hypertrophy of the anterior chamber

right ventricular hypertrophy, just as V3R may bring out the evidence of right ventricular hypertrophy in the normally placed heart V5R and V6R show the deep S waves of right ventricular dominance In spite of the frequency with which a right axis deviation occurs in patients with persistent cyanosis, a right axis deviation is extremely rare in cases of dextrocardia even in patients who show persistent cyanosis In Figure XXXIII-4 the standard leads show an extreme right axis deviation and the unipolar precordial leads show evidence of extreme 'right' ventricular hypertrophy

DIAGNOSIS

The condition can usually be diagnosed by careful physical examination The most significant single finding is the apex thrust in the fourth or fifth right inter

space at or beyond the mid-clavicular line This may be determined by inspection or palpation When the heart sounds are better heard to the right of the sternum than to the left, the existence of a dextrocardia should be suspected The diagnosis is definitively established by fluoroscopy or by a correctly taken electrocardiogram

DIFFERENTIAL DIAGNOSIS

A dextrocardia must always be differentiated from displacement of the heart and incomplete rotation of the heart upon its axis If the heart and the abdominal organs are the mirror image of the normal, the condition is clearly that of a situs inversus In contrast to this, in cases of displacement or incomplete rotation of the heart, the abdominal organs usually occupy their normal position but may be slightly displaced

Displacement of the heart usually occurs as a result of pulmonary pathology, such as atelectasis, pneumothorax, or massive pleural effusion Congenital absence or hypoplasia of the right lung with overexpansion of the left lung may displace the heart into the right thoracic cavity With the exception of a congenital anomaly of the lung (see Chapter XXII, Section c), displacement of the heart is usually the result of an acquired disease If, however, a patient with a congenital malformation of the heart develops pulmonary disease, this may cause displacement of the heart into the right thoracic cavity

Incomplete rotation of the heart on its axis, that is, dextroversion, is not always easy to differentiate from the complete reversal of the cardiac loop which occurs in a dextrocardia In a dextrocardia the apex beat is located at the lower, outer margin of the cardiac dullness In cases of incomplete rotation the apex thrust may lie close to the sternum, or well within the area of cardiac dullness

Although the clinical differentiation of a dextrocardia from marked displacement or incomplete rotation of the heart upon its axis may be extremely difficult, an electrocardiogram will usually differentiate the two conditions In a true dextrocardia the electrocardiogram shows characteristic changes (see Figure XXXIII-2), whereas in cases of a displacement or incomplete rotation of the heart the standard leads of the electrocardiogram remain essentially normal Even with extreme displacement or rotation, the P wave in Lead I remains upright, as does the main deflection of the ventricular complex, and usually the T wave in Lead I remains upright

If there is any evidence of an additional cardiac abnormality, the differentiation of a dextrocardia from an incomplete rotation of the heart upon its axis is

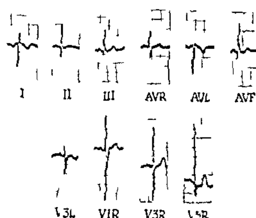


FIGURE XXXIII-3 Dextrocardia with dominance of the posterior chamber (unfortunately V-L was not taken)

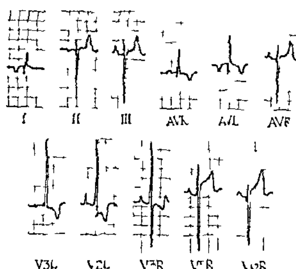


FIGURE XXXIII-4 Dextrocardia with hypertrophy of the anterior chamber

right ventricular hypertrophy, just as V_3R may bring out the evidence of right ventricular hypertrophy in the normally placed heart V R and V_6R show the deep S waves of right ventricular dominance In spite of the frequency with which a right axis deviation occurs in patients with persistent cyanosis, a right axis deviation is extremely rare in cases of dextrocardia even in patients who show persistent cyanosis In Figure XXXIII-4 the standard leads show an extreme right axis deviation and the unipolar precordial leads show evidence of extreme 'right' ventricular hypertrophy

DIAGNOSIS

The condition can usually be diagnosed by careful physical examination The most significant single finding is the apex thrust in the fourth or fifth right inter

B *Dextrorotation of the Heart*

Dextrorotation of the heart means that the primitive cardiac loop has rotated to the right The auricles occupy their normal position the abnormality primarily occurs in the bulboventricular loop The condition is almost invariably associated with some gross abnormality in the formation of the heart a single auricle is common and so is a functionally single ventricle, the great vessels also are frequently transposed A detailed anatomical diagnosis of the malformation is extremely difficult Nevertheless, if due consideration is given to all factors, it is usually possible to make a relatively accurate diagnosis on a broad functional basis

NATURE OF THE MALFORMATION

Dextrorotation of the heart means that the primitive cardiac loop swings to the right instead of to the left The abnormal rotation primarily concerns the ventricular loop, as the primitive cardiac tube is fixed at its venous end and at its arterial outlet Consequently, the position of the auricles is more constant than that of the ventricles and, although the great vessels may be rotated, they always arise from the base of the heart The left auricle usually lies posteriorly and the right auricle to the right of the sternum Generally the pulmonary veins open into the left auricle, and the venae cavae into the right auricle There may, however, be two superior venae cavae and occasionally the inferior vena cava is absent and the azygos vein pierces the diaphragm on the left and enters the left auricle at an abnormally high level or the hepatic vein may enter close to the pulmonary veins (see Figure xxxIII–5) Anomalies of the pulmonary veins may also occur Defects in the auricular septum are extremely common Grant[2] believes that a defect in the ventricular septum almost always occurs In the

Moreover, as the

vessels are rotated in such an abnormal manner that the pulmonary arteries and the aorta fail to meet the right and left ventricles in the normal manner Hence abnormal rotation of the great vessels is also the rule

Abnormalities of the other thoracic and abdominal viscera are common Grant[3] has emphasized the frequency with which a partial inversion of abdominal organs occurs with dextrorotation of the heart Ivemark[4] was the first to report the frequent occurrence of splenic agenesis and abnormalities of the gastro-intestinal tract with severe malformations of the heart

If the heart is rotated on its axis or the abdominal viscera are abnormally

important in diagnosis Only in cases of true *dextrocardia* is the heart the mirror image of the normal, in such instances alone is the assumption justified that the right ventricle lies anteriorly and the left ventricle posteriorly When this is true, the analysis of the additional malformation can be made on the same principle as when the heart occupies its normal position

When there is incomplete rotation of the heart, the assumption that the right ventricle lies anteriorly is no longer justified The electrocardiogram will indicate whether the anterior or the posterior chamber is enlarged, x ray and fluoroscopy will show whether the circulation to the lungs is increased or decreased The analysis of the abnormality is based on the principles outlined in Section B

TREATMENT

The dextrocardia per se calls for no special treatment When it occurs in combination with another cardiac malformation, treatment should be in accordance with that indicated for the associated malformation

PROGNOSIS

When a dextrocardia occurs in association with a situs inversus and without *any* evidence of a further malformation of the heart, the prognosis is excellent When a dextrocardia is combined with another malformation, the prognosis varies with the nature of the associated malformation

SUMMARY

A dextrocardia may occur with or without a situs inversus The diagnosis is established by careful physical examination, by fluoroscopy, or by an electrocardiogram

When a dextrocardia is combined with another cardiac malformation, the *clinical diagnosis* is complicated A dextrocardia combined with a malformation which causes persistent cyanosis is usually extremely severe and is difficult to diagnose clinically (see Section B) Such malformations are, however, frequently associated with a persistent left aortic arch Indeed, a left aortic arch in a dextrocardia combined with a malformation which causes persistent cyanosis is more common than a right aortic arch in a heart which occupies its normal position Interpretations of the oblique views are difficult because the apparently normal left aortic arch is associated with abnormal rotation of the ventricles The condition is asymptomatic and requires no treatment Prognosis is excellent, a true situs inversus is readily compatible with a normal active life

placed, careful consideration must always be given not only *to the intracardiac abnormalities* but also to the relation of the auricles to the ventricles and *the ventricles to the great vessels*

COURSE OF THE CIRCULATION

The course of the circulation varies with the structure of the heart An example of the course of the circulation in a patient with dextrorotation of the heart combined with a single auricle, a single ventricle, a *transposition* of the great vessels, and a small pulmonary artery is shown in Diagram XXXIII-1

PHYSIOLOGY OF THE MALFORMATION

The physiology of the malformation varies with the anatomical structure of the heart If there is complete admixture of venous and arterial blood within the auricles or the ventricles, the crucial factors are the size of the pulmonary artery and the pressure within that vessel If the pulmonary artery is of normal size and functionally there is a single ventricle, severe pulmonary hypertension is inevitable The ideal balance is to have a pulmonary stenosis severe enough to break the force of ejection from the common ventricle and yet not so severe as seriously to curtail the circulation to the lungs

CLINICAL FINDINGS

A good history is of real help Such a history will reveal the age at which cyanosis was first apparent, whether the infant suffered from attacks of paroxysmal dyspnea, whether the child squats when tired, and also the extent of his exercise tolerance Broadly speaking infants with pulmonary stenosis and a venous-arterial shunt suffer from attacks of paroxysmal dyspnea as the ductus arteriosus undergoes obliteration Consequently, if there is no history of paroxysmal dyspnea, either there is no venous-arterial shunt or the malformation is such that closure of the ductus arteriosus did not affect the circulation to the lungs This is generally true when there is but a single ventricle or a single arterial trunk

A slow gain in weight and *retardation of growth and development* in infancy are common when anoxemia is severe or when there is a large left-to-right shunt

Repeated respiratory and pulmonary infections are common when there is excessive pulmonary blood flow

Partial inversion of the abdominal organs is common [3] The *liver* frequently occupies a mid line position

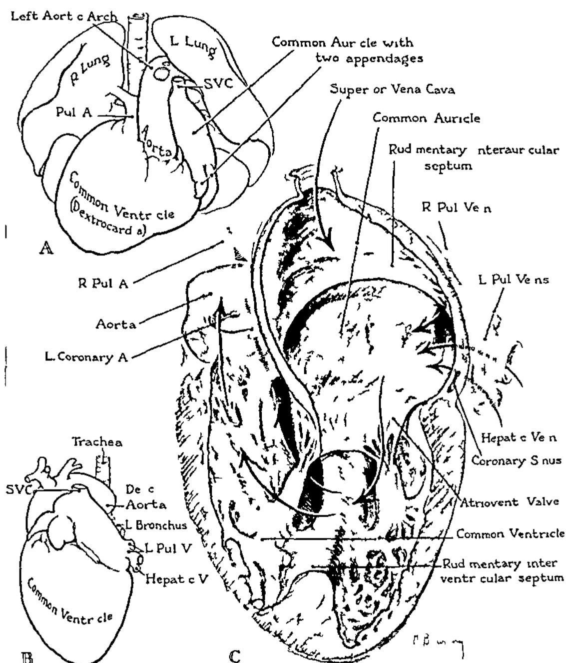


Figure xxxiii–5 Dextroversion with a single auricle a single ventricle and pulmonary stenosis Adult

DIAGRAM XXXIII-1

Dextrorotation combined with a single auricle a single ventricle transposition of the great vessels and a small pulmonary artery

In this malformation the heart is rotated to the right instead of to the left Such an abnormal rotation is usually associated with a serious cardiac abnormality Frequently there is a single auricle or a single ventricle with or without pulmonary stenosis and with or without transposition of the great vessels

In this instance a dextrorotation of the heart is combined with a single auricle, a single ventricle, transposition of the great vessels and an abnormally small pulmonary artery

All the blood from the superior and inferior venae cavae is directed into one side of the common auricle and that from the pulmonary veins into the other side of the common auricle The blood from the auricle flows through both the mitral and the tricuspid valves into the common ventricle Most of the blood from the common ventricle is pumped out into the aorta and systemic circulation This blood is returned to the heart in the normal fashion by the superior and inferior venae cavae. Only a small amount of the blood from the common ventricle is pumped through the small pulmonary orifice into the pulmonary artery The blood in the pulmonary artery flows to the lungs and is returned by the pulmonary veins to the left side of the common auricle Inasmuch as only a small volume of blood is pumped out into the pulmonary artery, a small volume of oxygenated blood is mixed with the large volume of venous blood returned by the superior and inferior venae cavae Cyanosis is intense.

Clinical diagnosis is made on a broad functional basis X ray or fluoroscopy reveals whether the pulmonary vascularity is markedly reduced, approximately normal, or markedly increased The electrocardiogram indicates whether the anterior or the posterior chamber is enlarged

A more definitive diagnosis usually requires angiocardiography and/or cardiac catheterization If the pulmonary blood flow is greatly reduced and the pressure in the pulmonary artery is low a systemic pulmonary anastomosis may benefit the patient

Diagram XXXIII-1

Head and upper extremities

R. Lung

L. Lung

RA

PV

Aorta

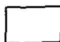
Common

Auricle

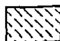
Common Ventricle

Right Descending Aorta

Trunk and lower extremities



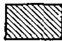
Arterial blood (fully saturated)



Small admixture of venous blood
No visible cyanosis



Venous and arterial blood
Cyanosis visible



Venous blood

right ventricle lies anterior to the left is not justified Nevertheless, it is usually possible to determine whether the anterior or the *posterior chamber* is enlarged and whether the pulmonary artery lies anterior or posterior to the aorta *In the* evaluation of the findings in the oblique positions, it is important to remember that when the heart has rotated to the right, the left and right anterior-oblique positions tend to replace each other Nevertheless, *if there is backward* displacement of the esophagus, it is usually due to enlargement of the left auricle

The relation of the esophagus to the aorta, however, depends upon whether the aorta arches to the right or whether there is a persistent left aortic arch

A left aortic arch in cases of dextrocardia combined with a malformation which causes persistent cyanosis is relatively common A left aortic arch bears the same relation to a dextrocardia as a right aortic arch does to the normal heart Just as a right aortic arch occurs most frequently in association with malformations of the great vessels, so a left aortic arch frequently *occurs in dextro*cardia in combination with a serious malformation which causes persistent cyanosis This is true regardless of whether or not there is a situs inversus

The position of the aorta is determined by x ray or fluoroscopic examination The diagnosis is not difficult In the anterior posterior position, if the aortic knob is visible *it is seen to lie to the left of the sternum and the esophagus is* displaced slightly to the right and indented upon its left, as shown in Figure XXXIII-6 The course of the esophagus in both oblique positions is entirely normal Thus in the right anterior-oblique position the aorta impinges upon the esophagus and slightly displaces it backward, and in the left anterior-oblique position the course of the esophagus is independent of the aorta, as *shown in* Figure XXXIII-7 A *left aortic arch is a familiar picture* and easy to recognize Indeed it is so familiar that both the clinician and the pathologist are likely to forget that a left aortic arch is abnormal in cases of dextrocardia

The difficult problem *is not the recognition* of the left aortic arch but the further analysis of the nature of the abnormality Although the aortic knob is visible on the left side and its relation to the esophagus appears *entirely normal,* the relation of the aorta to the pulmonary *artery and to the* ventricles is grossly altered *The pulmonary artery lies to the* right of the aorta and the great vessels cannot meet the ventricles in the normal fashion, as these are the mirror image of the normal The analysis of the nature of the malformation is extremely complicated because the changes in the aorta should be analyzed as if the heart were normal even though the changes in the cardiac contour are the mirror image of the normal For example, an abnormally clear *pulmonary* window is best visual

Abnormalities of the spleen are also common There may be accessory spleens or splenic agenesis Bush and Ainger showed that splenic agenesis can be diagnosed on the basis of hematological changes in the blood The condition has been considered by some to constitute a contraindication to surgery Although it is true that the abnormality is frequently associated with extremely complex cardiac abnormalities, in the author's opinion agenesis of the spleen is not a contraindication to surgery A systemic pulmonary anastomosis proved to be of great benefit to one patient who ten years later was found to have splenic agenesis

CARDIAC FINDINGS

The position and location of the apex thrust give an indication of the size and position of the heart At times the point of maximal impulse is clearly seen far to the right In other instances the cardiac beat can be felt equally well to the right and to the left These pulsations are not necessarily caused by the ventricles The strength of the pulsations which may be palpable over a greatly enlarged left auricle is truly remarkable Indeed, the author has studied two patients in whom the cardiac impulse was so vigorous in the fifth interspace in the left midclavicular line that it was difficult to convince others that there was dextrorotation and that the pulsations were transmitted from the left auricle, but such proved to be the case at autopsy

Murmurs and *thrills* are often of significance in the location of the heart, their variability, however, is as great as in other malformations

X RAY AND FLUOROSCOPIC FINDINGS

Fluoroscopic examination reveals the condition at a glance Once the existence of the dextrorotation is established, the positions of the liver and the stomach should be determined In the analysis of cardiovascular findings due consideration must be given to the abnormal position of the heart

It is usually relatively easy to determine whether the circulation to the lungs is approximately normal, greatly increased, or excessively reduced, and whether the aorta arches to the right or to the left

The auricles, as previously mentioned, are seldom grossly displaced The left auricle usually lies posteriorly and extends laterally to form the lower border of the cardiac shadow to the left of the sternum Although the right auricle usually lies to the right of the sternum, it is often displaced by the ventricles The outer margin of the cardiac silhouette to the right of the sternum is usually cast by the ventricle It is, however, important to remember that the assumption that the



Right anterior-oblique position



Left anterior-oblique position

FIGURE XXXIII-7 Dextrocardia with a left aortic arch Adult

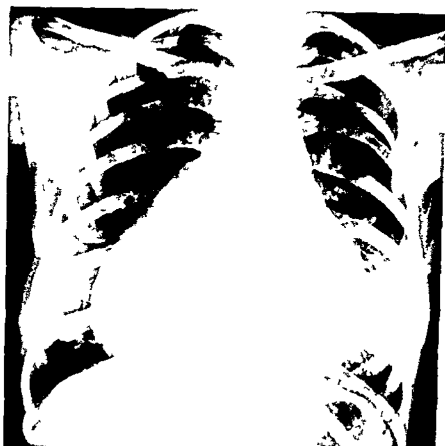


FIGURE XXXIII–6 Dextrocardia with a left aortic arch Adult

ized in the left anterior-oblique position, whereas the relative size of the two ventricles is determined in the right anterior oblique position Hence the analysis of abnormal cardiac shadows becomes extremely confusing

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram is of aid in the diagnosis The P waves are inverted in Lead I in a true dextrocardia and remain upright when there is dextrorotation of the heart Inversion of the P waves in Lead II and Lead III is usually indicative of an abnormal origin of the pacemaker

When the unipolar precordial leads are correctly placed over the right chest, they will offer evidence as to the relative thickness of the anterior and posterior portions of the ventricular musculature V L and V_3L record the deflections over the anterior chamber, and V_5R and V_6R those over the posterior chamber Thus a tall R wave and inversion of the T wave in V_2L and V_3L indicate hypertrophy of the anterior chamber, similar findings in V_5R and V_6R indicate dominance of the posterior chamber

made on a broad functional basis Thus it is usually possible to determine whether or not the patient suffers from reduced pulmonary blood flow or from pulmonary hypertension

DIFFERENTIAL DIAGNOSIS

Virtually the only differential diagnosis which is repeatedly brought to the fore is that of dextrocardia versus dextrorotation This differentiation is of importance in the analysis of the underlying malformation because in a true dextrocardia the left ventricle lies posteriorly and the right ventricle anteriorly, whereas in a dextrorotation this is no longer necessarily true If the P waves in Lead I are inverted and the apex impulse lies far to the right, the condition is probably a dextrocardia If the P waves are upright in Lead I, the condition is probably a dextrorotation or is caused by displacement of the heart to the right Angiocardiography may be necessary to determine the position of the right auricle

TREATMENT

Treatment should be conservative unless the patient is seriously incapacitated, as the underlying condition is usually more complicated than is apparent and the risk of operation is proportionally high Furthermore, there is always the possibility that the results of surgery may be unsatisfactory because of some unsuspected cardiac abnormality

If the patient suffers from a marked reduction in pulmonary blood flow, he can frequently be benefited by a Blalock Taussig operation or a Potts procedure

If, on the other hand, there is excessive circulation to the lungs and pulmonary hypertension, a pulmonary tuck, as recommended by Muller and Dammann[6] to break the pressure in the pulmonary artery and reduce the pulmonary blood flow, may be of great benefit to the patient

If total correction of the malformation is considered, extremely careful consideration must be given to the relation of the auricles to the ventricles, as well as of the ventricles to the great vessels because the nature of the malformation is often extraordinarily complex

PROGNOSIS

The four most important factors in the determination of the prognosis are whether the malformation is such as to cause progressive cardiac enlargement, whether blood is ejected into the lungs under systemic pressure, whether there is pulmonary stenosis and the degree of incapacity of the patient

SPECIAL TESTS

Hematology When the red blood cell count and the amount of available hemoglobin are at high normal levels, it usually indicates the existence of slight oxygen unsaturation of the arterial blood

Hematologic abnormalities similar to those which occur after splenectomy are found in patients with splenic agenesis [5] Thus the presence of target cells, normoblasts and Howell Jolly bodies in a blood smear suggests absence of the spleen In the author's experience splenic agenesis is not a contraindication to operation nor is its demonstration necessary to determine that the condition is complicated

The determination of the oxygen saturation of the arterial blood may be necessary to establish the presence or absence of a small right to-left shunt

Cardiac catheterization and the visualization of the course taken by the catheter give an indication of the size and position of the right auricle It may or may not be possible to enter the ventricle and only rarely is it possible to catheterize one of the great vessels Needless to say, the more complete the catheterization, the greater is the information obtained A word of caution concerning the findings in the great vessels the finding of systemic pressure in a great vessel in which the oxygen saturation of the blood is the same as that in the femoral artery does not prove that that vessel is the aorta, because if there is a single ventricle the pressure and the oxygen saturation in the pulmonary artery are the same as in the aorta The path which the catheter follows as it is advanced beyond the ventricle is frequently the decisive factor in the determination of whether the aorta or the pulmonary artery has been entered If the aorta has been entered, the catheter may be advanced into the neck vessels or seen to pass down the descending aorta below the diaphragm If the catheter is in the pulmonary artery, it may be advanced far out into the lungs and fully oxygenated blood may be obtained at the time a wedge pressure is taken

Angiocardiography gives valuable information up to the point of the first major abnormality Frequently there is a single auricle or a single ventricle and therefore only limited information is obtainable Usually angiocardiography does reveal the position and the course of both the aorta and the pulmonary artery, and it also offers a clue as to the size of the pulmonary vessels It will give definite evidence as to whether there is simultaneous opacification of the great vessels or which one is first delineated

DIAGNOSIS

When all factors are given due consideration, the diagnosis can usually be

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If the incapacity is slight and the increase in the size of the heart is directly proportional to the growth of the individual, the prognosis is good When these findings are combined with a normal pressure in the pulmonary artery and normal pulmonary blood flow, the prognosis is excellent

SUMMARY

Dextrorotation of the heart means that the primitive cardiac loop has in completely rotated to the right When the individual shows persistent cyanosis there is usually a gross anomaly of the heart The occurrence of a single auricle or a single ventricle is common Usually diagnosis can be made on a broad functional basis

The *history* is of help, especially as regards the age of the patient when cyanosis became apparent and whether or not he has suffered from paroxysmal dyspnea

Cardiac findings are variable The location of the apex thrust and the point of maximum intensity of the murmurs and thrills offer evidence as to the size and position of the heart

X ray and fluoroscopy show the size of the heart, the extent of the vascularity of the lungs, the position of the aortic arch, and whether or not the anterior or the posterior part of the ventricle is greatly enlarged

The electrocardiogram gives information as to the relative thickness of the anterior and posterior ventricular musculature

The determination of the oxygen saturation of the arterial blood may reveal oxygen unsaturation which is above the threshold of visible cyanosis

Cardiac catheterization will give information concerning the admixture of venous and arterial blood and concerning the pressure in the various chambers and the great vessels

Angiocardiography will reveal the position of the right auricle and will show whether both great vessels are simultaneously opacified

The diagnosis is made on a broad functional basis

Treatment varies with the nature of the malformation and the incapacity of the patient If there is reduced pulmonary blood flow, a Blalock-Taussig operation may be of great help If there is excessive circulation to the lungs, a pulmonary tuck may relieve the pulmonary hypertension and excessive pulmonary blood flow

If complete correction of the malformation is contemplated, the diagnosis must be checked with great care

The prognosis varies with the basic nature of the malformation

Young and Griswold,[4] in their report in 1951 of patients with a situs inversus and levocardia who had Blalock Taussig anastomoses at the Johns Hopkins Hospital, essentially confirmed the observations of Lochte and Forgacs Seven out of 8 of their cases showed evidence of anomalies of the systemic venous return and anomalies of the pulmonary venous return were demonstrated in 4 cases and suspected in another Defects in the auricular septum were found in 4 and suspected in 2 other instances In all 8 cases there was evidence of some degree of dextroposition of the aorta a complete transposition of the great vessels was demonstrated in 2 cases and postulated in 3 others

The first of the 8 cases in that report was the one which was studied by the author and originally misdiagnosed as a tetralogy of Fallot At autopsy the author noted that "the cardiac abnormality was most unusual, and commented as follows "The ventricle which structurally resembled the right ventricle lay anteriorly and that which resembled the left ventricle lay posteriorly, but the auricle which received the pulmonary veins opened into the anterior ventricle and that which received the superior and inferior venae cavae opened into the posterior ventricle Furthermore, the aorta arose from the posterior ventricle and partially over rode the ventricular septum and a small pulmonary artery arose from the anterior chamber The lungs, as well as the abdominal viscera, were reversed." In this case there was a transposition of the auricles but the great vessels were not transposed, thus instead of a corrected transposition there was in effect a transposition of the great vessels with pulmonary stenosis even though the aorta arose from the left ventricle

NATURE OF THE MALFORMATION

The basic nature of this malformation is that the auricles are transposed and the great vessels are also transposed A transposition of the auricles is usual when there is a situs inversus of the abdominal organs because under such circumstances the inferior vena cava lies to the left of the vertebral column Inasmuch as there are two superior venae cavae, it is natural for the one to persist which is on the side to which the venous blood from the trunk and the lower extremities is returned Hence, when the inferior vena cava lies on the left, the left superior vena cava persists

In a situs inversus with levocardia the heart rotates back to the left When the primitive cardiac loop swings back to the left, the bulboventricular loop is normally formed The right ventricle develops anteriorly from the bulbus cordis and the left ventricle forms posteriorly in the normal manner Under such cir

CHAPTER XXXIV

SITUS INVERSUS WITH LEVOCARDIA

TRANSPOSITION OF THE AURICLES

A SITUS INVERSUS of the abdominal viscera with a heart which occupies its normal position usually means that there is a complete reversal of all the organs and that the apex of the heart has rotated back to the left, hence the name a situs inversus with levocardia

When there is a situs inversus of the abdominal viscera, the position of the abdominal organs and blood vessels is the mirror image of the normal Hence the inferior vena cava lies on the left Furthermore, in a situs inversus it is normal for the left superior vena cava to persist The result is that the right, or caval, auricle lies on the left and the left,' or pulmonary, auricle tends to develop to the right of the caval auricle, in other words, the auricles are transposed

Whinnie[1] in 1840 described a case of situs inversus viscerum in which the inferior vena cava was on the left Lochte in 1898 reported the case of a cyanotic man with an incomplete rotation of the abdominal viscera in which he considered the auricles to be transposed He based his belief on the facts that there was a left superior vena cava and no *right* superior vena cava, that the inferior vena cava lay to the left of the vertebral column and entered the auricle which lay to the left, and, furthermore, that the inferior vena cava opened into the auricle close to the entrance of the coronary sinus

Maude Abbott was familiar with this type of corrected transposition The author remembers Dr Abbott's efforts to explain to her the meaning of a 'corrected transposition " Dr Abbott told her that the auricles were transposed, and so were the great vessels, and hence the one abnormality corrected the other At that time, however, the author did not fully understand the basic abnormality nor did she know that it occurred most commonly with a situs inversus of the abdominal viscera

In 1938 Forgacs[3] discussed the pathogenesis of cardiac defects in cases of isolated inversion of the abdominal viscera and emphasized that the development of the auricles was dependent on the position of the embryonic abdominal venous channels Hence a transposition of the auricles occurred when there was an isolated situs inversus of the abdominal organs or when there was an isolated dextrocardia

Young and Griswold,[4] in their report in 1951 of patients with a situs inversus and levocardia who had Blalock Taussig anastomoses at the Johns Hopkins Hospital, essentially confirmed the observations of Lochte and Forgacs Seven out of 8 of their cases showed evidence of anomalies of the systemic venous return and anomalies of the pulmonary venous return were demonstrated in 4 cases and suspected in another Defects in the auricular septum were found in 4 and suspected in 2 other instances In all 8 cases there was evidence of some degree of dextroposition of the aorta a complete transposition of the great vessels was demonstrated in 2 cases and postulated in 3 others

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NATURE OF THE MALFORMATION

The basic nature of this malformation is that the auricles are transposed and the great vessels are also transposed A transposition of the auricles is usual when there is a situs inversus of the abdominal organs, because under such circumstances the inferior vena cava lies to the left of the vertebral column Inasmuch as there are two superior venae cavae it is natural for the one to persist which is on the side to which the venous blood from the trunk and the lower extremities is returned Hence when the inferior vena cava lies on the left, the left superior vena cava persists

In a situs inversus with levocardia the heart rotates back to the left When the primitive cardiac loop swings back to the left, the bulboventricular loop is normally formed The right ventricle develops anteriorly from the bulbus cordis and the left ventricle forms posteriorly in the normal manner Under such cir

cumstances the "right," or caval, auricle opens into the left ventricle and left," or pulmonary, auricle opens into the right ventricle

The primitive cardiac loop is fixed at its arterial end as well as at its ven end Consequently the great vessels are also influenced by the visceral inversi Therefore, as the heart rotates back to the left, there is a tendency for the gr vessels to be transposed When this occurs, the aorta arises from the right ven cle to the left of and anterior to the pulmonary artery, and the pulmonary art arises from the left ventricle to the right of and posterior to the aorta Under su circumstances, although the "right" auricle opens into the left ventricle, the p monary artery arises from this ventricle, the 'left" auricle, which opens into t right ventricle, directs the blood to the aorta In other words, the auricles : transposed but in addition the great vessels are transposed, thus the transpositi of the one 'corrects" the transposition of the other

It is also possible for the inferior vena cava to be absent and the azygos s tem to develop as the main pathway for the return of venous blood from t lower extremities (see Chapter xxxii) Under such circumstances the veno blood from the abdomen is returned to the auricle on the right side When tl occurs it is common to find that the superior vena cava persists on the *right* a thus the venous blood is returned to the right auricle in the normal manner U der such circumstances, when the primitive cardiac loop swings to the left, tl normal forces are set in motion and the heart may develop normally except f the absence of the inferior vena cava

It is also possible for the inferior vena cava to cross over and pierce the di phragm in the normal manner If this happens, the venous auricle may readi develop in its normal position and the heart and circulation may be entire normal Nevertheless, a normal heart is unusual with a situs inversus and lev cardia

Cases of a situs inversus with levocardia in which the circulation was a parently normal have been reported [6] The author has seen one such patient wit a functionally normal heart, this patient had a normal right heart catheterizatio and a normal venous angiocardiogram, but the point of entrance of the inferic vena cava was not demonstrated In this instance the contour of the heart wa abnormal in that there was a high arch above the region of the normal pulmc nary conus (see Figures xxxiv–1 and 2)

COURSE OF THE CIRCULATION

The blood from the right auricle flows into the left ventricle and is pumpec

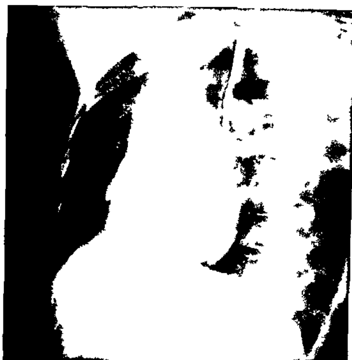


FIGURE XXXIV-1 Situs inversus with levocardia and a functionally normal heart Child

out through the pulmonary artery to the lungs, where it is oxygenated. The oxygenated blood is returned by the pulmonary veins to the left auricle (which lies to the right) and flows into the anatomical right ventricle (which lies anteriorly), thence it is pumped out through the aorta to the body The venous blood is returned by the inferior vena cava (which lies on the left) and by the left superior vena cava to the auricle which lies on the left but has the structure of the right auricle There the cycle starts again The course of the circulation is shown in Diagram XXXIV-1

PHYSIOLOGY OF THE MALFORMATION

The physiology of the malformation is normal when transposition of the auricles and the great vessels is complete As this seldom happens, additional malformations are common The manner in which the superimposed malformation alters the physiology varies with the nature of the abnormality The most important changes in the hemodynamics depend primarily on whether there is pulmonary stenosis or whether the lungs receive blood under systemic pressure



Left anterior oblique position



Right anterior oblique position

FIGURE XXXIV-2 Situs inversus with levocardia and a functionally normal heart Child

CLINICAL FINDINGS

Although it is possible for the patient to have a normal circulation, this is comparatively rare Furthermore, it is the occurrence of the additional malformation which usually brings the patient to the doctor

Cyanosis is a common complaint in patients with a *situs inversus and levocardia*, as malformations of the great vessels and the manner in which they meet the ventricles are common

The exercise tolerance of the individual varies with the oxygen saturation of the arterial blood and the patient s ability to increase his pulmonary blood flow with exercise Infants with a venous-arterial shunt and severe pulmonary stenosis may suffer from attacks of paroxysmal dyspnea and children with this condition will squat when tired

The outstanding finding of diagnostic importance is the position of the abdominal viscera

The liver lies on the left but accurate palpation of a liver of normal size is not easy If the liver is not palpable, the condition may be readily overlooked

The stomach lies on the right Indeed, the abdominal findings are identical with those in a complete situs inversus (see Chapter XXXIII, Section A)

The spleen may be abnormal there may be multiple spleens or splenic agenesis

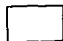
CARDIAC FINDINGS

The heart occupies its normal position The apex beat is palpable to the left The cardiac findings vary with the concomitant malformation but the apparent normality of the position of the heart renders diagnosis of the underlying abnormality difficult Indeed, the contour of the heart may closely resemble that of a familiar malformation and consequently the physician is *not alerted to the* true condition The basic nature of the abnormality is often first revealed by x ray or fluoroscopy

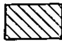
X RAY AND FLUOROSCOPIC FINDINGS

The heart occupies its normal position Its contour varies with the associated malformation but may appear so normal or so characteristic of a tetralogy of Fallot or some other malformation that the possibility of a more complicated condition is not considered until the abnormal position of the abdominal organs is seen Figure XXXIV–3 shows the x ray of the patient whose condition was diagnosed as a tetralogy of Fallot (see page 985) but who proved to have a transposi

Diagram XXXIV-1



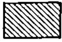
Arterial blood (fully saturated)



Venous and arterial blood
Cyanosis visible



Small admixture of venous blood
No visible cyanosis



Venous blood

DIAGRAM XXXIV-1

Situs inversus and levocardia (transposition of the auricles and transposition of the great vessels)

In this malformation there is an initial inversion of all the organs of the body. The liver lies on the left and stomach on the right. The heart is rotated back to the left. Nevertheless the caval auricle develops in relation to the superior vena cava and inferior vena cava. The inferior vena cava lies on the left; hence the right, or caval, auricle tends to lie on the left. The ventricular loop is, however, rotated normally to the left; hence the left ventricle lies posteriorly and the right ventricle lies anteriorly. The left or pulmonary auricle opens into the right ventricle and the caval auricle opens into the posterior left ventricle. The great vessels tend to be transposed. If the transposition is complete, the transposition of the great vessels corrects the transposition of the auricles and the basic course of the circulation is normal.

The blood from the right auricle flows into the left ventricle and is pumped out through the pulmonary artery to the lungs, where it is oxygenated. The oxygenated blood is returned by the pulmonary veins to the left auricle; thence it flows into the right ventricle and is pumped out through the aorta to the systemic circulation. The blood from the head and the upper extremities is returned by the left superior vena cava and that from the trunk and the lower extremities is returned by the inferior vena cava to the right auricle. There the cycle starts again.

Clinical diagnosis. The condition is to be suspected in a patient whose heart is in the normal position but who has an inversion of the abdominal organs. The condition is frequently complicated by additional abnormalities of the heart and great vessels. The diagnosis is extremely complicated. It is always necessary not only to consider the relation of the great vessels to the ventricles but also to study the relation of the auricles to the ventricles.



FIGURE XXXIX-3 Situs inversus and levocardia (same patient as in Figure XXXIX-4) Child

tion of the great vessels and a pulmonary stenosis In this instance the patient was too ill for fluoroscopy and the abnormal position of the abdominal viscera was not visible in the x ray This error in diagnosis clearly indicates that in every fluoroscopy it should be a routine procedure to lower the fluoroscopic screen after the administration of barium in order to determine the location of the stomach

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram may be entirely normal Although inversion of the P waves might be expected because of the abnormal position of the right auricle, such has not been our experience either when the heart was apparently normal or when the auricles were proven to be transposed The electrocardiogram of a patient with proven transposition of the auricles is shown in Figure XXXIX-4 Arrhythmias are extremely common

SPECIAL TESTS

Special tests are not needed Fluoroscopy or x ray after a barium swallow is the only study necessary to establish the fact that the abdominal viscera are transposed

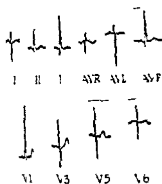


FIGURE XXXIV-4 Situs inversus and levo-*cardia* (same patient as in Figure XXXIV-3) Child

DIAGNOSIS

The diagnosis is based upon the *demonstration* of the abnormal position of the abdominal viscera in a patient whose heart occupies its normal position Once the basic diagnosis is made, detailed studies are necessary to determine the structure of the heart

When a patient with a situs inversus and levocardia presents himself for diagnosis of a malformation of the heart, analysis must be made with the realization that the auricles may be transposed, that the course of the inferior vena cava may be abnormal, and that the great vessels are probably transposed

DIFFERENTIAL DIAGNOSIS

There is none, as in no other condition are the abdominal viscera transposed and the heart in its normal position When the heart is apparently normal, special studies may be needed to confirm the fact and to determine the course of the inferior vena cava if this is of importance

Angiocardiography or cardiac catheterization performed through the saphenous vein will offer the best chance for the demonstration of the course of the venous return from the trunk and the lower extremities as well as the position of the right auricle (see Chapter XXXII)

COMMONLY ASSOCIATED MALFORMATIONS

Malformations of the great vessels and the manner in which they meet the ventricles are extremely common in patients with a situs inversus and levocardia

It is usually easy to determine whether there is adequate, excessive, or reduced pulmonary blood flow, and it is also easy to determine whether the aorta arches to the right or to the left and whether the anterior or posterior ventricle is hyper

trophied The relation of the great vessels to the ventricles should be studied in the usual manner In addition, it is essential to determine the position of the auricles in relation to the ventricles

Malformations of the auricular septum are also common An auricular septal defect should, however, never be closed unless it can be demonstrated that closure of the defect will give the individual a normal circulation The same precaution is essential prior to closure of a ventricular septal defect

TREATMENT

A corrected transposition of the auricles requires no treatment Nevertheless, the condition is frequently associated with some gross cardiac abnormality which causes persistent cyanosis

If the patient has reduced pulmonary blood flow and it is possible to demonstrate that venous blood is readily shunted into the aorta, he will probably be benefited by a systemic pulmonary anastomosis If, on the other hand, corrective surgery is contemplated, extremely accurate and detailed diagnosis is necessary The possibility of a transposition of the auricles as well as that of the great vessels must be borne in mind

PROGNOSIS

If the heart is functionally normal the prognosis is excellent When it is abnormal the prognosis varies with the nature of the malformation

SUMMARY

The basic malformation in a situs inversus with levocardia is a transposition of the auricles combined with a transposition of the great vessel, one abnormality compensates the other

When both abnormalities occur together, the circulation is basically normal and so is the physiology

The salient feature of the malformation is that the heart occupies its normal position and the abdominal viscera are the mirror image of the normal

The diagnosis is established by the demonstration of the abnormal position of the liver or the stomach Once the diagnosis is made, careful investigation is required to determine the course of the inferior vena cava and the position of the auricles Not only is it important to study the relation of the great vessels to the ventricles, but it is essential to know the relation of the auricles to the ventricles

If the patient has a normal circulation no treatment is necessary

If the patient suffers from reduced pulmonary blood flow, a systemic pulmonary anastomosis may help If corrective surgery is contemplated, extremely careful and detailed studies are necessary, as the diagnosis is seldom as simple as it appears and accurate diagnosis is essential

The prognosis depends upon the nature of the associated cardiac abnormality If the heart is functionally normal the prognosis is excellent

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CHAPTER XXXV

COMPLETE HEART BLOCK AND OTHER CARDIAC ARRHYTHMIAS

ALTHOUGH cardiac arrhythmias occur in association with congenital malformations of the heart, congenital complete heart block is virtually the only arrhythmia in which there is clear evidence that the condition is due to a congenital abnormality

A *Congenital Complete Heart Block*

Congenital complete heart block seldom occurs as an isolated abnormality Many types of cardiac malformations have been reported in association with it Septal defects, either auricular or of the high ventricular type, and corrected transpositions, are by far the *commonest of the malformations associated* with this conduction disturbance It is important to emphasize that auricular septal defects are quite as frequently associated with complete heart block as are defects in the ventricular septum The nature of the associated malformation is of no aid in the diagnosis of a complete dissociation The two conditions require separate analysis This section concerns only the diagnosis of complete heart block

NATURE OF THE MALFORMATION

A complete heart block means that there is complete interruption in the conduction of the impulses between the auricles and the ventricles Very few detailed studies[1 3] on the histology of the conduction system have been made, because of the tremendous work involved in the serial section of the conduction system For the latest reports on such studies, the reader is referred to the work of Lev[4] and his associates When complete dissociation is due to disease, it is usual to find an interruption in the main stem of the bundle of His between the atrioventricular node and its bifurcation into the two main branches The location of the defect is probably often in the same place when there is a congenital block This may explain why complete heart block so seldom occurs with the ventricular septal defects but may occur in association with defects low in the auricular septum and with a persistent ostium atrioventriculare commune

COURSE OF THE CIRCULATION

The complete interruption of the impulses between the auricles and the ventricles in no way alters the course of the circulation

PHYSIOLOGY OF THE MALFORMATION

Congenital complete dissociation does not fundamentally alter the physiology of the circulation It is, however, a striking fact that, although congenital complete heart block is readily compatible with life, heart block acquired during a cardiac operation is usually fatal Occasionally the child may live for months or for several years, but even after an apparently satisfactory adjustment sudden death may occur Why this type of heart block is so serious is not obvious, but that it is extremely serious is important to remember

CLINICAL FINDINGS

The condition is usually asymptomatic Stokes-Adams attacks seldom occur in childhood

CARDIAC FINDINGS

The cardiac findings in congenital complete heart block in no way differ from those in an acquired complete dissociation

The size of the heart is not appreciably altered by the complete dissociation (see Figure xxxv–1) The slow heart rate and the greater filling of the ventricles in diastole in all probability increase the size of the heart but the enlargement is so slight that the variation is within the limits of normality Indeed, the size of the heart, as well as the occurrence of thrills and murmurs, depends more upon the concomitant malformation than upon the complete dissociation

The slow regular heart rate offers the clue to the diagnosis It is, however, important to remember that the inherent cardiac rate varies not only with the size of the heart but also with the age of the individual The heart rate of an infant is normally faster than that of an adult and an infant with a complete heart block normally has a faster heart rate than does an adult with a similar abnormality

A complete dissociation with an idioventricular rate between 40 and 48 is generally readily compatible with life in an adult but in an infant such an excessively slow rate is of grave prognostic import. The author has seen several such infants, two died in the neonatal period and the others died suddenly within the first few months of life

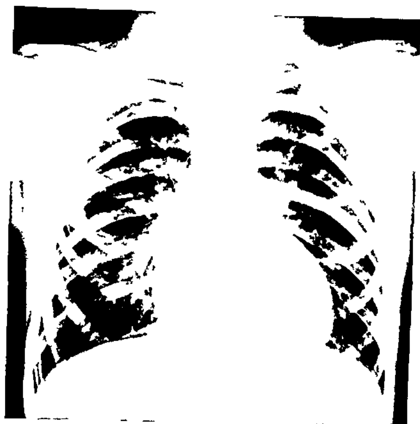


FIGURE XXXV-1 Congenital complete heart block Child

In contrast to this, when an infant is born with an idioventricular rate of approximately 80 per minute, the condition is usually compatible with a normal life During childhood the inherent cardiac rate gradually slows For this reason, in young children a slow regular rhythm with a rate of under 70 per minute should suggest the possibility of a complete dissociation

The absence of a sinus arrhythmia in a person with a slow heart rate is always suggestive of a complete dissociation Its absence in young individuals is especially significant because a sinus arrhythmia is the rule in childhood and, even when absent, it can usually be elicited by exercise or by deep breathing The demonstration of such an arrhythmia is positive proof that the individual does not have a complete heart block Conversely, the absence of a sinus arrhythmia in a child with a bradycardia should arouse suspicion of the possibility of a complete dissociation

A third heart sound which, though not constant, is clearly heard during the long diastolic pauses may be of aid in diagnosis The sound has the quality of a normal third heart sound In all probability, it is produced by the flow of blood from the auricles to the ventricles When this occurs in mid diastole, a sound is

clearly audible When the auricular contraction occurs close to the time of ventricular systole, the flow of blood from the auricle to the ventricle is inaudible For this reason the sound is inconstant The fact that there is a loud, but inconstant, sound in mid-diastole is suggestive of a complete dissociation

The character of the venous pulse in the neck vessels is often of aid in diagnosis Theoretically, the auricular waves can be seen to be independent of those caused by the contraction of the ventricles Although these waves can be recorded on a polygraphic tracing, they are difficult for the eye to discern The feature of clinical importance is the inequality in the strength of these pulsations in the vessels of the neck A series of small waves is interrupted by one or two conspicuous pulsations Such variation in the strength of the venous pulsations is suggestive of a complete heart block

The effect of exercise is also important Although in adults exercise usually has no effect upon the idioventricular rate, it is important to emphasize that in children exercise usually causes an acceleration of 10 to 20 beats per minute After exercise the heart rate slows down with a striking absence of a sinus arrhythmia

The effect of atropine is closely similar to that of exercise In adults with complete heart block, atropine, even in large doses, seldom alters the idioventricular rate In children atropine commonly causes an increase in both the auricular and the ventricular rate, the complete dissociation, however, persists

X RAY AND FLUOROSCOPIC FINDINGS

The heart usually appears slightly enlarged The cardiac contractions are so vigorous that they cause a significant change in the size of the heart in systole and diastole

The contour of the heart is, however determined by the concomitant malformation

ELECTROCARDIOGRAPHIC FINDINGS

The electrocardiogram offers positive proof of the presence or absence of a complete heart block (see Figure xxxv–2) The electrocardiogram in no way differs from that of an acquired complete heart block The ventricular rate is regular and is slower than the auricular rate The P waves also are regular and are seen to fall in all phases of the cardiac cycle For detailed analysis of the electrocardiographic findings the reader is referred to the authorities on electrocardiography

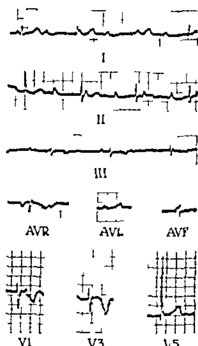


FIGURE XXXI-2 Congenital complete heart block

DIAGNOSIS

The condition should be suspected when there is a slow, regular heart rate and it is impossible to elicit a sinus arrhythmia The occurrence of a distinct, though inconstant, heart sound in mid-diastole may offer an additional clue The variation in the strength of the venous pulse in the neck vessels is also suggestive of a complete heart block The diagnosis is established by the finding of a complete dissociation in the electrocardiogram The proof that the condition is congenital depends partly upon the age at which the heart block was first detected and partly upon the other findings in the heart (see under Differential Diagnosis)

DIFFERENTIAL DIAGNOSIS

The condition is to be differentiated from other bradycardias, from a partial heart block, and from acquired complete heart block

A sinus bradycardia may be extreme It is usually accompanied by a marked sinus arrhythmia The presence of a sinus arrhythmia always excludes the possibility of a complete dissociation In the absence of a sinus arrhythmia, an electrocardiogram may be necessary to differentiate the two conditions

A partial heart block only occasionally simulates a complete heart block Simple prolongation of the conduction time does not alter the heart rate Dropped beats usually produce an irregular rhythm A partial heart block is confused

with a complete dissociation only in cases in which there is a regular 2 to 1 or 3 to 1 block This type of block is extremely rare It has been reported in Mongolian idiots in combination with a persistent ostium atrioventriculare commune The diagnosis is proven by the electrocardiogram

Acquired complete heart block may be extremely difficult to differentiate from a congenital complete block The age at which the slow heart rate is first noted is of help Only if the condition has been detected at birth is it certain that the heart block is congenital in origin Nevertheless, inasmuch as the average practitioner pays little attention to the heart rate of a child, no great weight should be placed upon the fact that the condition was not discovered until late childhood or early adult life In cases of a congenital complete heart block it is common to find some evidence of an additional cardiac abnormality The occurrence of a murmur, however, does not necessarily mean that there is a congenital malformation of the heart Murmurs are the rule, not the exception, in severe rheumatic heart disease

Acute rheumatic fever is probably the most common cause of acquired heart block in children and young adults

Diphtheria was formerly regarded as a common cause of complete heart block Thompson, Golden, and White[5] have shown that most patients who survive a diphtheritic myocarditis make a complete recovery Furthermore, in the author s experience complete dissociation in diphtheria is almost always a terminal event In the rare cases in which recovery has occurred a normal sinus mechanism has been restored Therefore it is the author s belief that diphtheria rarely, if ever, is the cause of a complete heart block in an otherwise healthy individual

An auricular septal defect with a superimposed rheumatic infection is a possibility which should never be forgotten This combination of conditions may be associated with all forms of cardiac arrhythmias The signs of a congenital malformation of the heart may be so pronounced as to mask the signs of a rheumatic infection Therefore in all cases in which there is evidence of an auricular septal defect a complete dissociation should not be regarded as congenital in origin unless the occurrence of a superimposed rheumatic infection has been carefully excluded

TREATMENT

Complete heart block with a slow idioventricular rate of 40 to 50 per minute at birth is so serious that an effort should be made to increase the heart rate It

is possible that the use of a pacemaker might lead to the establishment of a more rapid idioventricular rate Therefore, just as soon as the condition is detected, such treatment seems worthy of trial

If at birth the idioventricular rate is 80 or above, the condition usually requires no treatment

If an older patient develops Stokes-Adams attacks, the treatment is the same as that for a patient with an acquired complete heart block

PROGNOSIS

The prognosis is usually excellent Only in infants with an exceptionally slow heart rate is the prognosis guarded The condition is usually compatible with a long and active life

SUMMARY

Congenital complete heart block may occur as an isolated malformation but usually occurs in association with some other malformation of the heart, such as a defect low in the auricular septum, a persistent ostium atrioventriculare commune, or a corrected transposition

The complete interruption of the conduction of impulses from the auricles to the ventricles causes a slow, regular idioventricular heart rate The heart rate is slower in adults than in younger individuals A heart rate of 80 per minute in an infant or below 70 in a child should arouse suspicion of the possibility of a complete heart block

The characteristic findings are the same as in other cases of complete dissociation The heart rate is slow and regular and there is often an inconstant third heart sound in mid diastole The venous pulse in the neck frequently shows a variation in the strength of the successive waves Exercise or atropine may cause an acceleration of both the auricular and the ventricular rate but has no influence upon the block

The diagnosis is established by the electrocardiogram The condition is usually asymptomatic and requires no treatment For infants born with an excessively slow heart rate, the prognosis is extremely grave For those born with an idioventricular rate of 80 per minute the prognosis is excellent

B *Other Cardiac Arrhythmias*

The Wolff Parkinson White syndrome of a short P-R interval and a prolongation of the QRS complex is generally believed to be caused by a congenital

anomaly in the conduction system Persons with this conduction disturbance *frequently suffer* from repeated attacks of paroxysmal tachycardia Such attacks should be treated in the usual manner Because of the predilection to paroxysmal tachycardia, infants with this conduction disturbance should be maintained on digitalis until sufficiently old so that the danger of severe decompensation from paroxysmal tachycardia is past (see below and also Chapter v)

Partial heart block that is, the prolongation of *the atrioventricular conduction time* with or without dropped beats, seldom occurs as an isolated congenital abnormality in an otherwise normal heart Indeed, in the presence of such a finding, a systemic infection, rheumatic or otherwise, must always be *excluded*

Prolongation of the P-R interval does, however, occur in certain malformations of the heart It is the rule in Ebstein s anomaly of the tricuspid valve and also frequently occurs in corrected transpositions In the latter group of malformations the condition may progress to dropped beats and even to complete dissociation

Slight prolongation of the atrioventricular conduction time is also common in a patent ductus arteriosus and in an auricular septal defect In the latter condition the possibility of a superimposed rheumatic infection must always be considered

Dropped beats are usually due to acquired heart disease A regular 2 to 1 heart block has been reported in patients with congenital malformations of the heart[8]

With advancing years some patients with congenital malformations of the heart, notably those with a defect in the auricular septum or a corrected *transposition* develop dropped beats There is no clear evidence whether this condition is the result of a congenital malformation or of a superimposed infection It seems probable that the conduction of impulses may be more readily *interfered* with if the bundle of His is displaced than if the heart is normal Nevertheless, the disturbance in rhythm is probably not caused by a congenital anomaly but by a sclerosing process associated with advancing years

Prolongation of the intraventricular conduction is, of course, the rule in a Wolff Parkinson White syndrome In glycogen storage disease of the heart the QRS interval is slightly prolonged in relation to the short P-R interval Slight prolongation of the QRS interval frequently *occurs in* an auricular septal defect of the ostium secundum type and is the rule in a defect *of the* ostium primum type Prolongation of the intraventricular conduction time may occur *and* progress in a corrected transposition and also when an aneurysm in the sinus of

Valsalva burrows through the ventricular septum and ruptures into the right ventricle

Extrasystoles, paroxysmal tachycardia, auricular flutter, and *auricular fibrillation* have all been reported at an early age Excessively slow, excessively rapid, and irregular heart action have been detected in utero [10] Such rhythms are clearly abnormal It does not, however, follow that they are produced by any abnormality in the structure of the heart Most of these arrhythmias are transitory, thus proving that the structure of the myocardium is fundamentally normal Even when the arrhythmia is persistent, inasmuch as the normal anatomical and histological structure of the conduction mechanism in the auricles is not known, it is impossible even at autopsy to prove that the arrhythmia was due to a congenital malformation of the auricular musculature Whether or not these arrhythmias are true congenital anomalies, they may develop during intrauterine life and are abnormal

Broadly speaking, cardiac arrhythmias are relatively common in cases of enormous enlargement of one or both auricles For example, cardiac arrhythmias of all sorts are common in patients who have auricular septal defects, anomalies of the venous return in which all the pulmonary veins drain into the right auricle, or Ebstein s anomaly

The etiology is obscure but, in all probability, most of these arrhythmias are caused by some extrinsic factor A complete discussion of cardiac arrhythmia is beyond the scope of this book The following discussion is confined to the arrhythmias which occur in early infancy, as the development of an arrhythmia later in life proves that the irregularity is not congenital in origin For a general discussion of arrhythmias the reader is referred to the authorities on electrocardiography and acquired heart disease

Extrasystoles have been recorded in the neonatal period, moreover, irregularities have been noted in utero which, at birth, were proven to be extrasystoles This does not, however, necessarily mean that the extrasystoles are due to a structural abnormality, nor does it mean that the arrhythmia will continue throughout life Extrasystoles are not usually associated with malformations of the heart, furthermore, if the heart is normal, extrasystoles in the neonatal period are of no greater significance than are those which occur later

Paroxysmal tachycardia although it occurs in infancy, has never been known to be congenital in origin The heart is usually entirely normal but the arrhythmia may be associated with a Wolff Parkinson White syndrome

The occurrence of paroxysmal tachycardia, although relatively benign in adult life, may be extremely serious in early infancy If the paroxysm continues *for many hours, the infant is in danger of* developing decompensation and may die of cardiac failure Indeed, the condition probably represents one of the explanations of the so-called idiopathic hypertrophy, that is, cardiac enlargement and cardiac failure of unknown etiology in which at autopsy nothing is found to explain the large heart

Prompt treatment is always indicated *Carotid sinus pressure* may be tried, *ocular pressure* if attempted, must be performed with care because of the danger of injury to the infant s eyes

Digitalis or one of the allied preparations is usually indicated because of the danger of cardiac failure The use of such preparations usually brings the paroxysm to an end Since a rapid effect is desired, either lanatoside C or digoxin is usually the drug of choice The dose should always be in direct proportion to the weight of the infant (for detailed treatment see Chapter x) The establishment of a normal sinus mechanism proves that the arrhythmia was not due to any inherent abnormality in the myocardium

Auricular flutter dating from intra uterine life is very rare but may occasionally occur in an infant with a congenital malformation of the heart Even in such a case, it is extremely doubtful whether the auricular flutter itself is due to a congenital anomaly in the auricular musculature in the sense that the structure of the auricle is such that a normal sinus mechanism can never be established Only once has the author seen a baby with an auricular flutter in whom the arrhythmia had been noted in intra uterine life In that instance the arrhythmia persisted for over seven months Nevertheless, between the ages of seven months and one year a normal sinus mechanism was established which proved that the auricular flutter was not caused by a congenital abnormality in the conduction system The child, however did have clear evidence of a congenital malformation of the heart

Auricular fibrillation may occur in infancy but to the author s knowledge no case has been reported in which it was known to date from intra uterine life Furthermore failure to establish a normal mechanism with digitalis or other appropriate therapy does not necessarily mean that there is no sinus node So little is known of the histology of the sinus node or of the normal conduction of impulses through the auricles that even at autopsy it is impossible to determine whether the arrhythmia was caused by an abnormality in the conduction mech

anism or was the result of some toxin or of damage to the auricular musculature Certainly in the vast majority of cases auricular fibrillation is acquired and not congenital in origin

The clinical manifestations, cardiac findings, and *treatment* of these arrhythmias are the same for a person with a congenital malformation of the heart as for all other patients For a complete discussion of this subject the reader is referred to the authorities on electrocardiography and acquired heart disease

SUMMARY

Partial heart block may occur in Ebstein's anomaly of the tricuspid valve and in corrected transpositions Such a heart block requires no treatment

Other cardiac arrhythmias also occur in patients with malformations of the heart The arrhythmia may be present at birth but it usually does not persist throughout life The establishment of a normal sinus rhythm proves that the arrhythmia is not due to a congenital abnormality in the conduction system Paroxysmal tachycardia may, however, occur in association with a Wolff Parkinson-White syndrome

The treatment of these arrhythmias is the same as for those which occur in any other patient, with the single exception of paroxysmal tachycardia This condition calls for prompt treatment with digitalis or some allied preparation, as there is real danger that an infant with paroxysmal tachycardia may develop cardiac failure and die Prompt treatment is especially important because, as soon as the paroxysm is brought to an end, the prognosis is excellent

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8. Dippel A. L. Two cases of congenital heart disease in which the diagnosis was made before birth Am J Obst & Gynec 27 120-123 1934

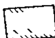
9. Sampson J J, R L. McCalla, and W M J Kerr Phonocardiography of the human fetus Am. Heart J 1 717-734 1926

10 Geiger C J and L. E Hines Prenatal diagnosis of complete congenital heart block JAMA 115 2272-2273, 1940

VISUAL INDEX



Arterial blood (fully aturated)

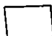


Small admixture f ven u l lood
N visible cy n s s



Venous and arterial bl od
Cyanosis ible

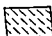
KEY TO PLATES 1–6



A terial blood (fully saturated)



Venous and arterial bl l
Cyanosis vi ible

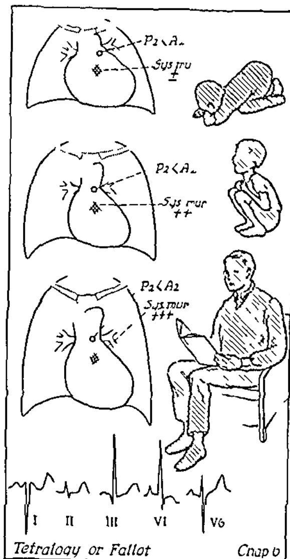


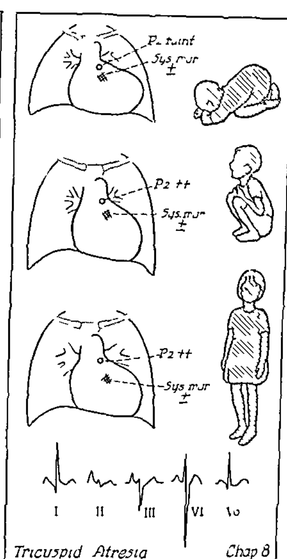
Small adm tur of   nou blood
N vis ble cyanosis

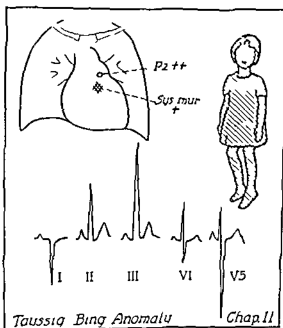


Ven us blood

KEY TO PLATES 7–8







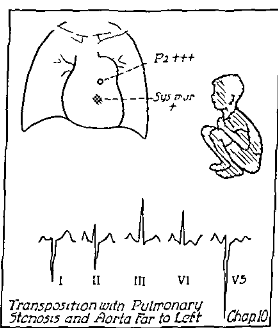
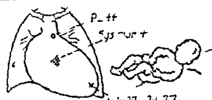
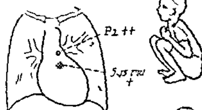
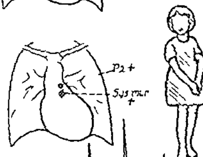
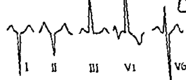


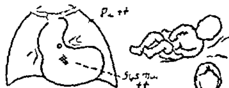
Plate 1 Malformation in which the shunt is from right to left, hence there is persistent cyanosis

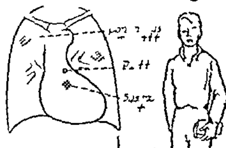
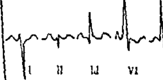


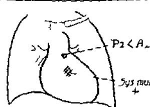


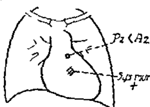


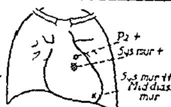




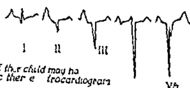













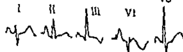
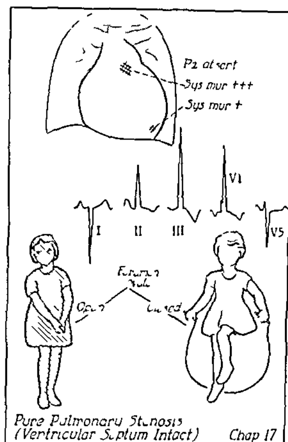
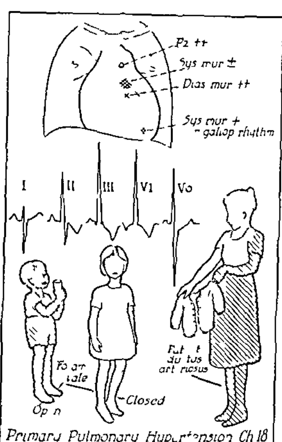




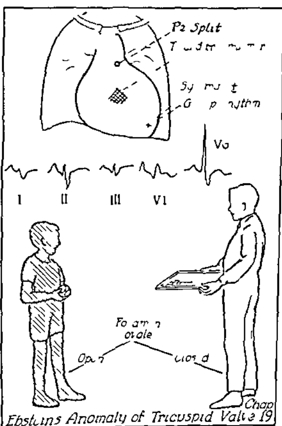
PLATE 2 Malformation in which the shunt is predominantly from right to left hence there is usually persistent cyanosis



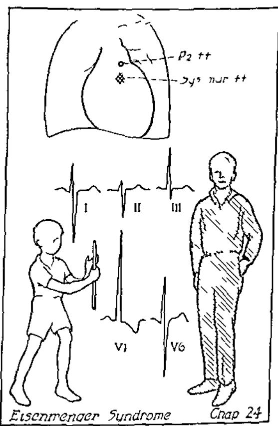
Pure Pulmonary Stenosis (Ventricular Septum Intact) Chap 17



Primary Pulmonary Hypertension Ch 18



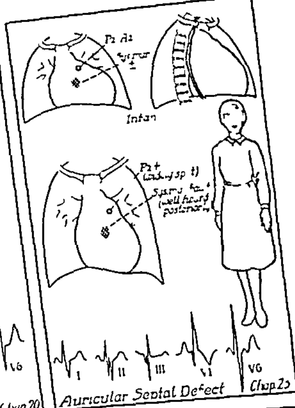
Ebsteins Anomaly of Tricuspid Valve Chap 19

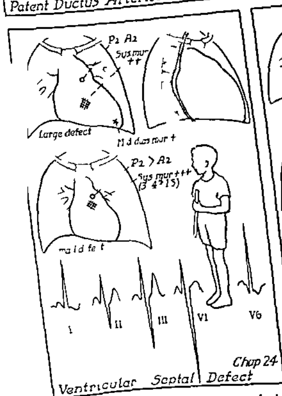


Eisenmenger Syndrome Chap 24

PLATE 3 Malformation in which there may be a right to left shunt, hence there may be cyanosis







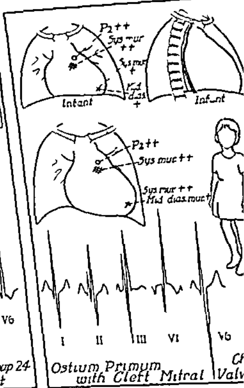


PLATE 4 Malformation in which the shunt is usually from left to right hence there is no cyanosis

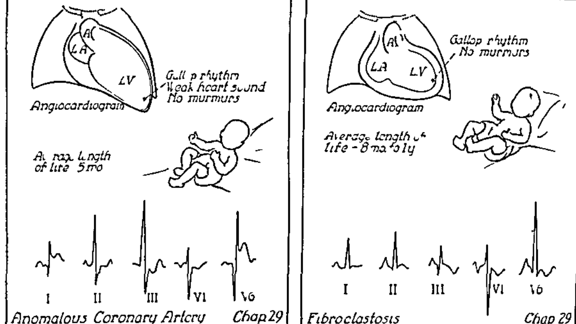


Plate 5a Malformation in which there is no shunt, hence no cyanosis

P2 +

Sys mur +

All pulmonary veins to coronary sinus

Faint cont. mur

Sys mur +

All pulmonary veins to right superior vena cava via left superior vena cava

P2 ++

Sys mur +

Sys mur ±

Partial anomaly of pulmonary venous return

Gallop rhythm

All pulmonary veins to inferior vena cava via hepatic vein

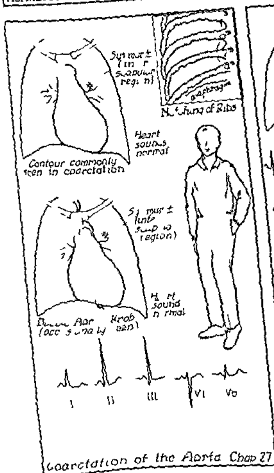
Average length of life - 21 days

I II III VI V6

Anomalies of Pulmonary Venous Return — Chap 22

Plate 5b Malformation in which the shunt is variable, usually there is no visible cyanosis

Normal Aortic Arch and Common Anomalies (Esophagrams)



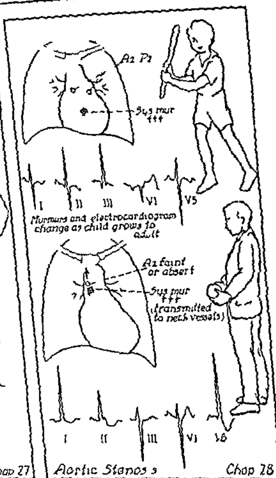
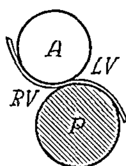
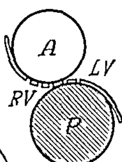


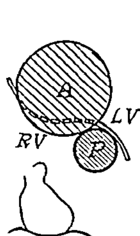
PLATE 6 Malformation in which there is no shunt and hence no cyanosis



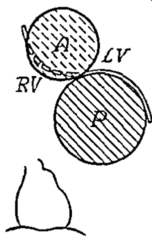
Normal



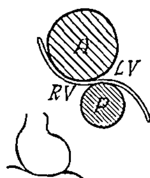
Ventricular septal defect



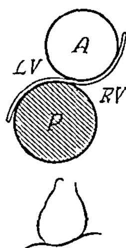
Tetralogy of Fallot



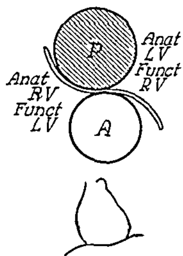
Eisenmenger complex



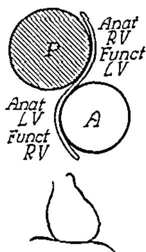
Defective development of right ventricle



Situs Inversus

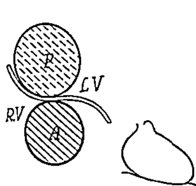


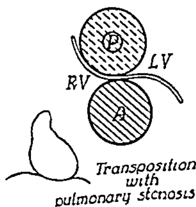
Situs Inversus with levocardia

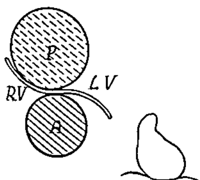


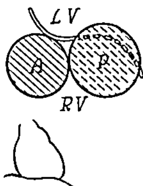
Corrected transposition

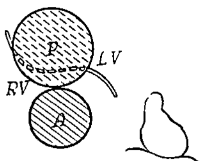
PLATE 7 Relation of the great vessels to the ventricular septum, pulmonary artery arising from the right ventricle











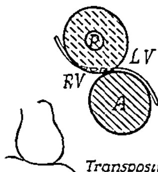
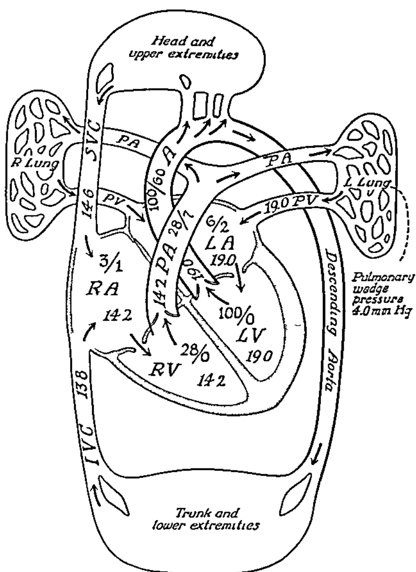


PLATE 8 Relation of the great vessels to the ventricular septum aorta arising from the right ventricle



Oxygen content expressed in Vol %
Blood pressure expressed in mm Hg

PLATE 9 Oxygen content and pressures in the normal heart

INDEX TO VOLUME II

INDEX TO VOLUME II

Note Cross references beginning with capital letters refer to main entries cross references beginning with small letters refer to sub-entries

E

E

P

Y

Z